**CONGENITAL HEART DISEASES AND OTHER VASCULAR CONDITIONS**

Septal Defects

Tetralogy of Fallot

Patent Ductus Arteriosus

**Cardiovascular conditions;** Hypertensive heart disease

Hypertensive crisis

Pulmonary heart disease

Heart murmurs

Myocardial infarction

Rheumatic heart disease

Hypertensive heart failure

Transposition of the great arteries

Tricuspid atresia

Hypoplastic left heart syndrome

Double outlet right ventricle

Pulmonary valve stenosis

Bicuspid aortic valve

Total anomalous pulmonary

Single ventricle defect

Coarctation aorta

Truncus arteriosus

**Septal defects (atrial septal defect, ventricular septal defect [ASD/VSD])**

**SEPTAL DEFECTS**

**{DEFINITION}**

Normally, in babies that do not have heart defects, the right side of the heart pumps blood that is poor in oxygen from the heart to the lungs, while the left side of the heart pumps blood that is rich in oxygen to the rest of the body.

Septal defects are commonly referred to as "**holes in the heart**." This term is used to describe congenital heart defects (that is, defects that are present at birth) where there is an abnormal opening in the walls (septa) that separate the left and the right sides of the heart's chambers.

**TYPES OF SEPTAL DEFECTS**

Specifically, there are two types of septal defects: ***Ventricular Septal Defects (VSDs)*** and ***Atrial Septal Defects (ASDs)*** are types of septal defects. VSDs involve a hole in the wall between the heart's lower chambers (ventricles), while ASDs involve a hole in the wall between the heart's upper chambers (atria).

**VENTRICULAR SEPTAL DEFECT (VSD)**

**Definition:** A hole in the wall that separates the heart's lower chambers (ventricles). In babies with a **VSD,** blood flows from the left ventricle through the **VSD** to the right ventricle into the lungs.

A ventricular septal defect (VSD) changes how blood flows through the heart and lungs. Oxygen-rich blood gets pumped back to the lungs instead of out to the body. The oxygen-rich blood mixes with the oxygen-poor blood. These changes may increase blood pressure in the lungs and require the heart to work harder to pump blood.

A small ventricular septal defect may cause no problems. Many small ventricular septal defects (VSDs) close on their own. Babies with medium or larger VSDs may need surgery in life to prevent complications.

Occurrence: About 42 of every 10,000 babies in the United States are born with a VSD. This means that about 16,800 babies are born with a VSD each year.

**Types of VSDs**

An infant with a VSD can have one or more holes in different places of the septum. There are several names for these holes. Some common locations and names are listed below:

**Conoventricular Ventricular Septal Defect:**In general, this is a hole where portions of the ventricular septum should meet just below the pulmonary and aortic valves.

**Perimembranous Ventricular Septal Defect:**This is a hole in the upper section of the ventricular septum.

**Inlet Ventricular Septal Defect:**This is a hole in the septum near to where the blood enters the ventricles through the tricuspid and mitral valves. This type of ventricular septal defect also might be part of another heart defect called an atrioventricular septal defect (AVSD).

**Muscular Ventricular Septal Defect:**This is a hole in the lower, muscular part of the ventricular septum. This is the most common type of ventricular septal defect.

**Causes of VSD**

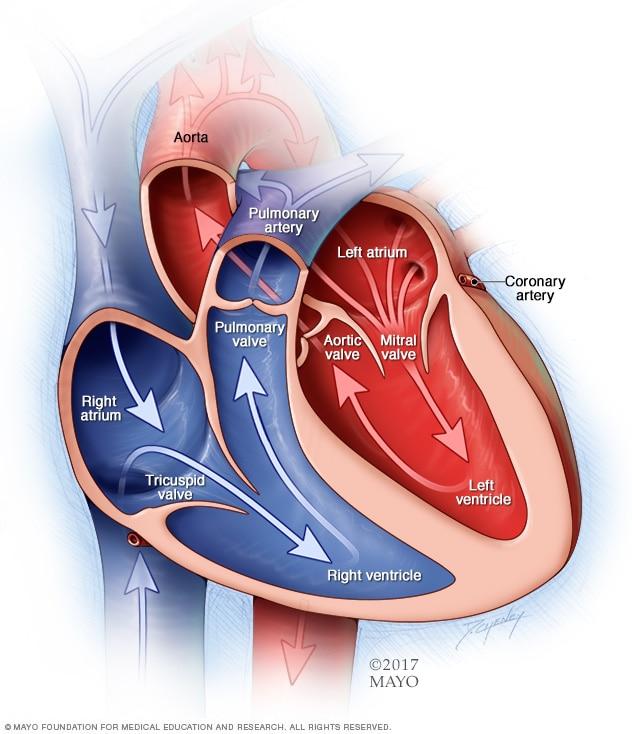
This usually happens during pregnancy if the wall that forms between the two ventricles does not fully develop; this then leaves a hole.

Ventricular septal defect (VSD) occurs as the baby's heart is developing during pregnancy. The muscular wall separating the heart into left and right sides doesn't form fully, leaving one or more holes. The size of the hole or holes can vary.

There's often no clear cause. Genetics and environmental factors may play a role. VSDs can occur alone or with other heart problems present at birth. Rarely, a ventricular septal defect can occur later in life after a heart attack or certain heart procedures.

**How the heart works**

To understand more about ventricular septal defect (VSD), it may be helpful to know how the heart typically works.



**Chambers and valves of the heart**

A typical heart has two upper and two lower chambers. The upper chambers, the right and left atria, receive incoming blood. The lower chambers, the more muscular right and left ventricles, pump blood out of the heart. The heart valves, which keep blood flowing in the right direction, are gates at the chamber openings.

The typical heart is made of four chambers — two upper chambers (atria) and two lower chambers (ventricles).

* The right side of the heart moves blood to the lungs.
* In the lungs, blood picks up oxygen.
* The lungs pump the oxygen-rich blood to the heart's left side.
* The left side of the heart pumps the oxygen-rich blood to the rest of the body.

A ventricular septal defect changes the direction of blood flow in the heart and lungs. The hole lets oxygen-rich blood go back into the lungs, instead of going out to the body. Oxygen-rich blood and oxygen-poor blood now mix together. If the ventricular septal defect is large, the blood pressure in the lung arteries may increase. The heart then must work harder to pump blood. A large VSD can also increase the amount of blood flow in the lung arteries, causing congestion.

**Risk Factors**

The causes of ventricular septal defects among most babies are as of yet still unknown. Some babies have heart defects because of changes in their genes or chromosomes. A combination of genes and other risk factors may increase the risk for ventricular septal defects. These factors include things in a mother’s environment, what she eats or drinks, or the medicines she uses.

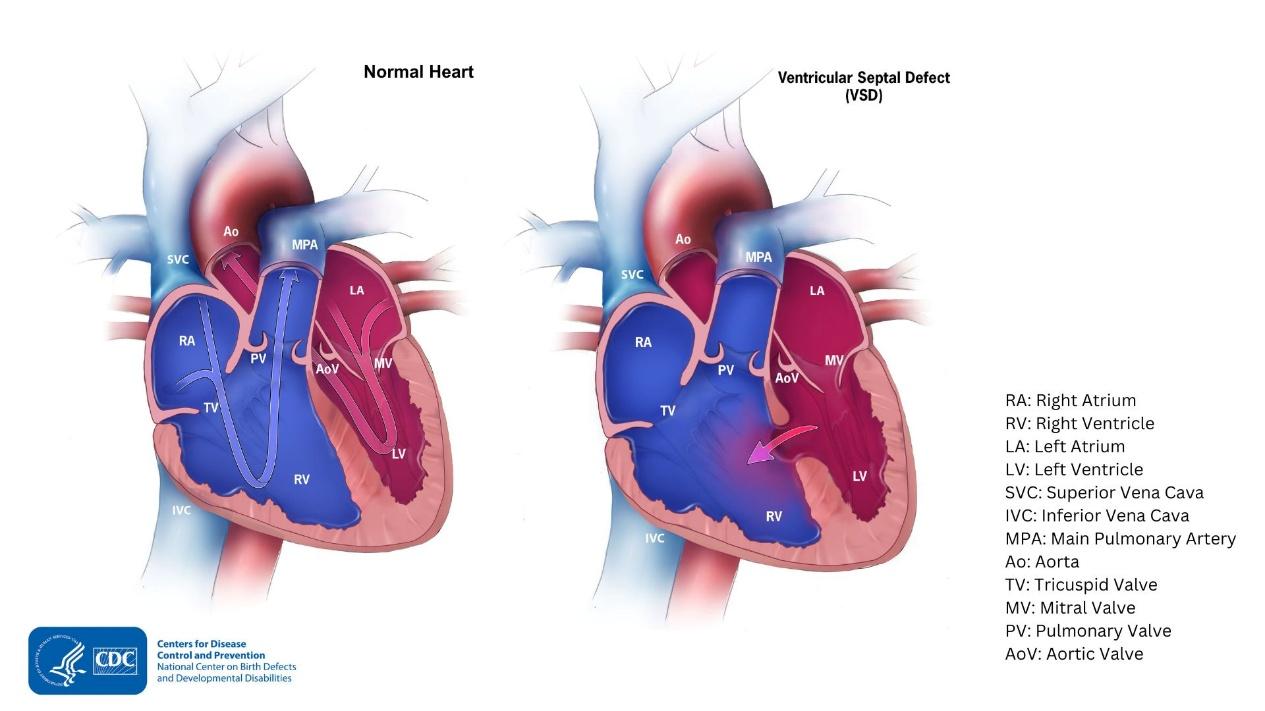
Risk factors for ventricular septal defect (VSDs) could also include:

* Premature birth
* Down syndrome and other genetic conditions
* Family history of heart problems presents at birth (congenital heart defects)

A baby born with ventricular septal defect may have other heart problems, such as:

* Atrial septal defect
* Coarctation of the aorta
* Double outlet syndrome
* Patent ductus arteriosus
* Tetralogy of Fallot

If you already have a child with a congenital heart defect, a genetic counselor can discuss the risk of your next child having one.



**Signs and symptoms**

The size of the ventricular septal defect will influence what symptoms, if any, are present.

Signs of a ventricular septal defect (VSD) might be present at birth, or might not appear until well after birth. If the hole is small, it could close on its own. The baby might not show any signs of the defect. However, if the hole is large, the baby might have symptoms, including:

* Shortness of breath
* Fast, heavy breathing or breathlessness
* Sweating
* Tiredness while feeding
* Poor weight gain
* Poor eating
* Slow or no physical growth (failure to thrive)
* Easily gets tired
* Enlarged liver
* Pale skin
* Whooshing sound when listening to the heart with a stethoscope (heart murmur)

Symptoms can occur a bit differently in each child. The symptoms of VSD may also be similar to symptoms of other conditions. Make sure your child sees the healthcare provider for a correct diagnosis.

Symptoms of a ventricular septal defect in adults may include:

* Shortness of breath, especially when exercising
* Whooshing sound when listening to the heart with a stethoscope (heart murmur)

**Diagnosis**

A VSD is usually diagnosed after a baby is born. During a physical examination, a healthcare provider might hear a whooshing sound called a heart murmur while listening to the heart with a stethoscope. The size of the VSD will influence whether a healthcare provider hears a heart murmur during a physical examination.

Some ventricular septal defects (VSDs) are diagnosed soon after a child is born. However, ventricular septal defects (VSDs) may not be diagnosed until later in life. Sometimes a ventricular septal defect (VSDs) can be detected by a pregnancy ultrasound before the baby is born.

Here below are a list of tests that can be done to help in diagnosing ventricular septal defects. These include:

**Echocardiogram:** This is one of the tests that are most commonly used to diagnose a ventricular septal defect. Sound waves are used to create pictures of the heart in motion. An echocardiogram can show how well blood is moving through the heart and heart valves.

**Electrocardiogram (ECG):** This quick and painless test records the electrical activity of the heart. It can show how fast or how slowly the heart is beating.

**Chest X-ray:** A chest X-ray shows the condition of the heart and lungs. It can tell if the heart is enlarged and if the lungs have extra fluid.

**Pulse oximetry:** A sensor placed on the fingertip records the amount of oxygen in the blood. Too little oxygen may be a sign of a heart or lung problem.

**Cardiac catheterization:** In this test, a thin, flexible tube (catheter) is inserted into a blood vessel at the groin or arm and guided through the blood vessels into the heart. Through cardiac catheterization, doctors can diagnose congenital heart defects and determine the function of the heart valves and chambers.

**Cardiac magnetic resonance imaging (MRI) scan:** Magnetic fields and radio waves are used to create detailed images of the heart. A health care provider might request this test if more information is needed after an echocardiogram.

**Computerized tomography (CT) scan:** A series of X-rays create detailed images of the heart. It may be done if an echocardiogram didn't provide as much information as needed.

**Treatment options**

Ventricular septal defect treatment may include regular health checkups, medications and surgery. Many babies born with a small ventricular septal defect (VSD) won't need surgery to close the hole. Some small VSDs close on their own.

If the VSD is small, regular health checkups may be all that's needed. Medication may be prescribed to treat any symptoms.

Babies who have large VSDs or who tire easily during feeding may need extra nutrition to help them grow. Some babies may require medication to help treat heart failure symptoms.

**Medications**

Medications won't repair a ventricular septal defect, but they may be given to treat symptoms or complications. The specific medications used depend on the symptoms and their cause. Water pills (diuretics) are used to decrease the amount of fluid in the body and reduce the strain on the heart.

Oxygen may be given.

Some babies and children will need medicines to help:

* Strengthen the heart muscle
* Lower their blood pressure
* Help the body get rid of extra fluid

**Nutrition**

Some babies with a ventricular septal defect become tired while feeding and do not eat enough to gain weight. To make sure babies have a healthy weight gain, a special high-calorie formula might be prescribed. Some babies become extremely tired while feeding and might need to be fed through a feeding tube.

**Surgeries or other procedures**

Surgery may be done if the VSD is medium or large or if it's causing severe symptoms. Babies who need surgery to repair the hole often have the procedure in their first year.

A surgeon may close small ventricular septal defects if their location in the heart could cause damage to nearby structures, such as the heart valves.

Surgeries and procedures to repair a ventricular septal defect include:

* **Open-heart surgery.** This is the preferred procedure for repairing most ventricular septal defects. A surgeon uses a patch or stitches to close the hole between the lower heart chambers. This type of VSD surgery requires a heart-lung machine and an incision in the chest.
* **Catheter procedure.** Some ventricular septal defects can be repaired using thin, flexible tubes (catheters) without the need for open-heart surgery. The health care provider inserts a catheter into a blood vessel, usually in the groin, and guides it to the heart. A small device is inserted through the catheter to close the hole.

After ventricular septal defect surgery, regular checkups are needed for life, ideally by a heart doctor (cardiologist). Checkups often include imaging tests to determine how well surgery is working.

**Prevention tips**

Because the cause is unclear, it may not be possible to prevent ventricular septal defect (VSD). But getting good prenatal care is important. If you have a VSD and are planning to become pregnant, schedule a visit with your health care provider and follow these steps:

* Get early prenatal care, even before you're pregnant. Talk to your provider before you get pregnant about your health and discuss any lifestyle changes that your doctor may recommend for a healthy pregnancy. Also, be sure you talk to your doctor about any medications you're taking.
* Take a multivitamin with folic acid. Taking 400 micrograms of folic acid daily has been shown to reduce birth defects in the brain and spinal cord. It may help reduce the risk of heart defects as well.
* Avoid alcohol. Drinking alcohol during pregnancy increases the risk of congenital heart defects.
* Don't smoke or use illegal drugs. If you smoke, quit. Smoking during pregnancy increases the risk of a congenital heart defect in the baby. Avoid using illegal drugs as they may harm a developing baby.
* Get recommended vaccinations. Be sure you're up to date on all of your vaccinations before becoming pregnant. Some infections can be harmful to a developing fetus. For example, having rubella (German measles) during pregnancy can cause problems in a baby's heart development. A blood test done before pregnancy can determine if you're immune to rubella. A vaccine is available for those who aren't immune.
* Keep diabetes under control. Careful control of blood sugar before and during pregnancy can reduce the risk of congenital heart defects in the baby. Diabetes that develops during pregnancy (gestational diabetes) generally doesn't increase a baby's risk. If you have diabetes, work with your provider to be sure it's well controlled before getting pregnant.
* Check with your provider before taking any medications. Some medications can cause birth defects. Tell your provider about all the medications you take, including those bought without a prescription.

If you have a family history of heart problems present at birth, consider talking with a genetic counselor and a heart doctor (cardiologist) before getting pregnant.

**Self-care**

Lifestyle changes may be recommended to keep the heart healthy and prevent complications.

* Prevent heart infections. Sometimes heart problems can increase the risk of infection in the lining of the heart or heart valves (endocarditis). Antibiotics may be recommended before dental procedures if you have low oxygen due to a large VSD. The medicines may also be recommended if you have a surgically repaired VSD with a patch that still has some blood flow across it. Antibiotics may also be recommended if you recently had catheter-based VSD repair.

For most people with a ventricular septal defect, good oral hygiene and regular dental checkups can prevent endocarditis.

* Ask about exercise restrictions. Many people with a ventricular septal defect can lead healthy, active lives without restrictions. But some may need to limit exercise and sports activities. Ask your health care provider which sports and types of exercise are safe for you or your child. People with Eisenmenger syndrome should avoid strenuous physical activity.
* Talk to your provider before getting pregnant. If you have a ventricular septal defect and are pregnant or hoping to be, talk to your health care provider about the possible risks and complications. Together you can discuss and plan for any special care needed during pregnancy.

A small VSD or a repaired one without complications doesn't pose a large additional pregnancy risk. However, a large, unrepaired VSD, irregular heart rhythms, heart failure or pulmonary hypertension increase the risk for pregnancy complications.

Pregnancy is considered a very high risk for those with Eisenmenger syndrome and is not recommended.

**Prognosis {Probable outcome or likelihood of recovery}**

The prognosis for a ventricular septal defect (VSD) largely depends on the size of the defect. Children with small VSDs are typically asymptomatic (meaning they do not show any signs or symptoms of VSDs) and have an excellent long-term prognosis, with no need for medical or surgical therapy. These defects often close spontaneously as the child grows, and regular checkups are usually sufficient to monitor the condition.

For moderate or large VSDs, medical therapy may be necessary to manage symptoms of congestive heart failure (CHF). Some VSDs may become smaller with time, but uncontrolled CHF symptoms with growth failure indicate the need for surgical repair. The long-term prognosis for repaired VSDs is generally good, with patients unlikely to have significant long-term problems if the defect is repaired early in life.

Patients with small VSDs that remain open have a small risk of developing endocarditis, a heart infection, and should maintain good oral hygiene. Additionally, a small nut definite risk of malignant ventricular arrhythmia exists, particularly for small Perimembranous VSDs.

In cases where a VSD does not close spontaneously and is large, the risk of developing pulmonary hypertension increases, which can lead to Eisenmenger syndrome if left untreated. This condition is associated with cyanosis and is considered very high risk for pregnancy.

Regular follow-ups with a cardiologist are essential for monitoring the condition and determining the appropriate course of action.

**Possible complications**

A small ventricular septal defect (VSD) may never cause any problems. Some medium or large VSDs may be life-threatening. Treatment can help prevent many complications.

Complications of ventricular septal defect can include:

* **Heart failure.** In a heart with a medium or large VSD, the heart works harder and the lungs have too much blood pumped to them. Without treatment, heart failure can develop.
* **Eisenmenger syndrome.** An unrepaired hole in the heart can lead to this complication after many years. Irregular blood flow causes the blood vessels in the lungs to become stiff and narrow. Blood pressure rises in the lungs' arteries (pulmonary hypertension). This syndrome permanently damages the blood vessels in the lungs.
* **Endocarditis.** This is a rare complication of VSD. An infection causes life-threatening inflammation of the inner lining of the heart's chambers and valves.
* **Other heart problems.** These include heart valve disease and irregular heart rhythms (arrhythmias).
* **Lung problems**
* **Irregular heart rhythms (arrhythmias)**
* **Heart valve problems**
* **Poor growth and complications**

**When to see a doctor / red flags**

You know it is time to see a doctor when you start noticing these symptoms; if your baby:

* Tires easily when eating or playing
* Is not gaining weight
* Becomes breathless when eating or crying
* Breathes rapidly or is short of breath

Call your healthcare provider if these symptoms develop:

* Shortness of breath
* Rapid or irregular heartbeat
* Fatigue or weakness

REFERENCE

[**https://www.mayoclinic.org/diseases-conditions/ventricular-septal-defect/symptoms-causes/syc-20353495**](https://www.mayoclinic.org/diseases-conditions/ventricular-septal-defect/symptoms-causes/syc-20353495)

[**https://www.cdc.gov/heart-defects/about/ventricular-septal-defect.html**](https://www.cdc.gov/heart-defects/about/ventricular-septal-defect.html)

**Atrial Septal Defect (ASD)**

A hole in the wall that separates the heart's upper chambers (atria).

**Definition/Description**

An atrial septal defect (ASD) is a heart condition that you’re born with. That means it’s a congenital heart defect. People with an ASD have a hole on the wall that separates between the left and right side of the upper heart chambers. The hole increases the amount of blood going through the lungs. It can vary in size and may close on its own or require surgery.

As a baby's heart develops during pregnancy, several openings in the wall divide the upper chambers of the heart (atria). These openings normally close during pregnancy or shortly after birth. If one of these openings does not close, a hole remains, and it is called an atrial septal defect (ASD).

Small atrial septal defects might be found by chance and never cause a concern. Others might close during infancy or early childhood.

A large, long-term atrial septal defect can damage the heart and lungs. Surgery may be needed to repair an atrial septal defect and to prevent complications.

**Occurrence:** About 13 of every 10,000 babies in the United States are born with an atrial septal defect.This means that about 5,240 babies are born with an atrial septal defect each year.

**Types**

The types of atrial septal defects (ASDs) include:

* Secundum. This is the most common type of ASD. It occurs in the middle of the wall between the upper heart chambers. This wall is called the atrial septum.
* Primum. This type of ASD affects the lower part of the wall between the upper heart chambers. It might occur with other heart problems present at birth.
* Sinus venosus. This is a rare type of ASD. It most often happens in the upper part of the wall between the heart chambers. It often occurs with other heart structure changes present at birth.
* Coronary sinus. The coronary sinus is part of the vein system of the heart. In this rare type of ASD, part of the wall between the coronary sinus and the left upper heart chamber is missing.

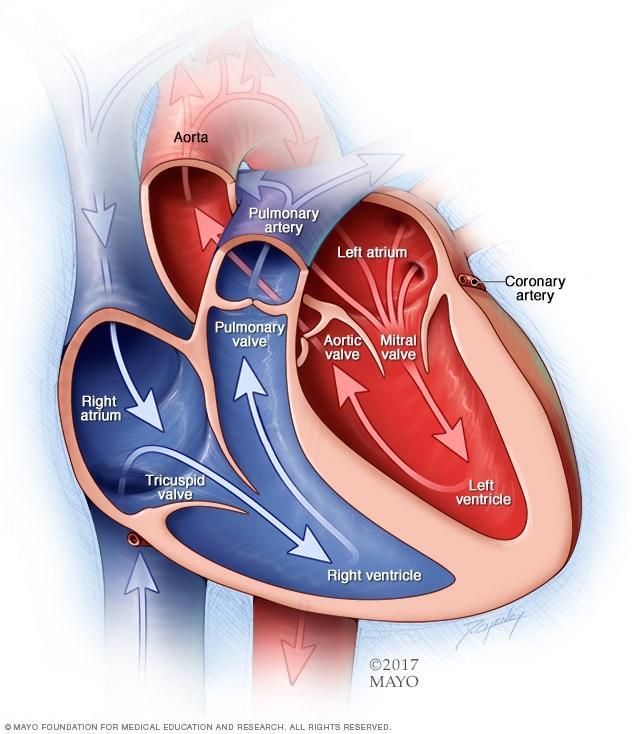
**Causes**

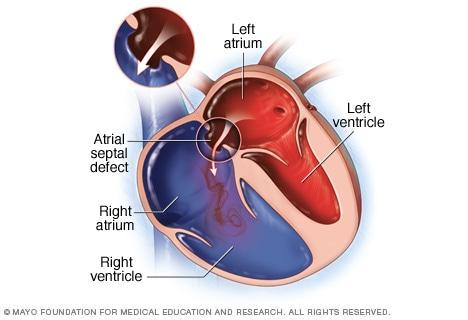
The cause of atrial septal defect is not clear. The problem affects the structure of the heart. It happens as the baby's heart is forming during pregnancy.

The following may play a role in the cause of congenital heart defects such as atrial septal defect:

* Changes in genes.
* Some medical conditions.
* Certain medicines.
* Smoking.
* Alcohol misuse.

**How the heart works**

 **Chambers and valves of the heart**

 Atrial Septal Defect (ASD)

To understand the cause of atrial septal defect, it may be helpful to know how the heart typically works.

The typical heart is made of four chambers. The two upper chambers are called the atria. The two lower chambers are called the ventricles.

The right side of the heart moves blood to the lungs. In the lungs, blood picks up oxygen and then returns it to the heart's left side. The left side of the heart then pumps the blood through the body's main artery, called the aorta. The blood then goes out to the rest of the body.

A large atrial septal defect can send extra blood to the lungs and cause the right side of the heart to work too hard. Without treatment, the right side of the heart grows larger over time and becomes weak. The blood pressure in the arteries in the lungs also can increase, causing pulmonary hypertension.

**Risk factors**

The causes of atrial septal defects among most babies are unknown. Some babies have heart defects because of changes in their genes or chromosomes. A combination of genes and other risk factors may increase the risk for atrial septal defects. These factors can include things in a mother's environment, what she eats or drinks, or the medicines she uses.

Atrial septal defect (ASD) occurs as the baby's heart is forming during pregnancy. It is a congenital heart defect. Things that may increase a baby's risk of atrial septal defect or other heart problems present at birth include:

* German measles, also called rubella, during the first few months of pregnancy.
* Diabetes.
* Lupus.
* Alcohol or tobacco use during pregnancy.
* Cocaine usage during pregnancy.
* Use of some medicines during pregnancy, including those to treat seizures and mood conditions.

Some types of congenital heart defects occur in families. This means they are inherited. Tell your care team if you or someone in your family had a heart problem present at birth. Screening by a genetic counselor can help show the risk of certain heart defects in future children.

**Signs and symptoms**

An atrial septal defect is usually present at birth, but most babies born with an atrial septal defect (ASD) may not have any signs or symptoms. Symptoms may begin in adulthood. The known signs and symptoms of a large or untreated septal defect may include:

Atrial septal defect symptoms may include:

* Difficulty breathing or shortness of breath, especially when exercising.
* Tiredness, especially with activity.
* Swelling of the legs, feet or belly area.
* Irregular heartbeats, also called arrhythmias.
* Skipped heartbeats or feelings of a quick, pounding or fluttering heartbeat, called palpitations.
* A whooshing sound that can be heard with a stethoscope (heart murmur)
* Stroke

**Diagnosis methods**

Some atrial septal defects (ASDs) are found before or soon after a child is born. But smaller ones may not be found until later in life.

If an ASD is present, a healthcare professional may hear a whooshing sound called a heart murmur when listening to the heart with a device called a stethoscope.

An atrial septal defect may be diagnosed during pregnancy or after the baby is born.

**During pregnancy**

During pregnancy, screening tests (prenatal tests) check for birth defects and other conditions. An ultrasound, a tool that creates pictures of the baby, may detect an atrial septal defect. However, it usually depends on the size of the hole and its location. If an atrial septal defect is suspected, a healthcare provider will need to confirm the diagnosis.

**After a baby is born**

In many cases, an atrial septal defect may not be diagnosed until adulthood. An atrial septal defect is often found by detecting a murmur when listening to a person's heart with a stethoscope. If a murmur is heard or other symptoms are present, the healthcare provider might request more tests to confirm the diagnosis. The most common test is an echocardiogram, which is an ultrasound of the heart.

If an ASD is present, a healthcare professional may hear a whooshing sound called a heart murmur when listening to the heart with a device called a stethoscope.

**Tests:** Some tests that can be used to diagnose if an atrial septal defect (ASD) is present include:

* Echocardiogram. This is the main test used to diagnose an atrial septal defect. Sound waves are used to make pictures of the beating heart. An echocardiogram shows the structure of the heart chambers and valves. It also shows how well blood moves through the heart and heart valves.
* Chest X-ray. A chest X-ray shows the condition of the heart and lungs.
* Electrocardiogram (ECG or EKG). This quick and painless test records the electrical activity of the heart. It can show how fast or how slow the heart is beating. An ECG can help find irregular heartbeats, called arrhythmias.
* Cardiac magnetic resonance imaging (MRI) scan. This imaging test uses magnetic fields and radio waves to make detailed images of the heart. It might be done if other tests didn't provide a sure diagnosis.
* Computerized tomography (CT) scan. This test uses a series of X-rays to create detailed pictures of the heart. It may be used if other tests don't give enough information to make a diagnosis.

**Treatment options**

Treatment for atrial septal defect (ASD) depends on many factors:

* The age when the diagnosis is done
* The size of the hole in the heart.
* The amount of seriousness of symptoms
* Whether there are conditions or heart problems present at birth.

An atrial septal defect may close on its own during childhood. For small holes that don't close, regular health checkups may be the only care needed.

Some atrial septal defects that do not close need a procedure to close the hole. But closure of an ASD isn't recommended in those who have severe pulmonary hypertension.

Sometimes surgery is needed to repair the hole. Medications can be prescribed to help treat symptoms. There are no known medications that can repair the hole.

With an atrial septal defect diagnosis, the healthcare provider may monitor it to see if the hole closes on its own. During this period of time, the healthcare provider might treat symptoms with medicine. If a child has a large atrial septal defect, the healthcare provider may recommend that it be closed. This will prevent problems later in life.

Closure may also be recommended for an adult who has many or severe symptoms. Closure of the hole may be done during cardiac catheterization or open-heart surgery. After these procedures, follow-up care will depend on a few factors:

* The size of the defect
* The person's age
* Whether the person has other birth defects

Sometimes the atrial septal defect can't be fully repaired. However, procedures to close the hole can improve blood flow and the way the heart works. For some people, even if their heart defect has been repaired, they are not cured and will require follow-up care.

**Medications**

Medicines won't repair an atrial septal defect (ASD). But they can help reduce symptoms. Medicines for atrial septal defect might include:

* Beta blockers to control the heartbeat.
* Blood thinners, called anticoagulants, to lower the risk of blood clots.
* Diuretics to reduce fluid buildup in the lungs and other parts of the body.

**Surgery or other procedures**

A procedure is often suggested to repair a medium to large atrial septal defect (ASD) to prevent future complications.

Atrial septal defect repair involves closing the hole in the heart. This can be done two ways:

* **Catheter-based repair.** This type is done to fix the secundum type of atrial septal defects. A thin, flexible tube called a catheter is put into a blood vessel, most often in the groin. The tube is then guided to the heart. A mesh patch or plug goes through the catheter. The patch is used to close the hole. Heart tissue grows around the patch, closing the hole for life. However, some large secundum atrial septal defects might need open-heart surgery.
* **Open-heart surgery.** This type of ASD repair surgery involves making a cut through the chest wall to get to the heart. The surgeons use patches to close the hole. Open-heart repair surgery is the only way to fix primum, sinus venosus and coronary sinus atrial defects.

Sometimes, atrial septal defect repair can be done using smaller cuts than traditional surgery. This method is called minimally invasive surgery. If the repair is done with the help of a robot, it's called robot-assisted heart surgery.

Anyone who has had surgery for atrial septal defect needs regular imaging tests and health checkups. These appointments are to watch for possible heart and lung complications.

People with large atrial septal defects who do not have surgery to close the hole often have worse long-term outcomes. They may have more trouble doing everyday activities. This is called reduced functional capacity. They also are at greater risk for irregular heartbeats and pulmonary hypertension.

**Lifestyle and home remedies**

Following a heart-healthy lifestyle is important. This includes eating healthy, not smoking, managing weight and getting enough sleep. If you or your child has an atrial septal defect, talk to your healthcare team about the following:

* **Exercise.** Exercise is usually OK for people with an atrial septal defect. But if ASD repair is needed, you might have to stop certain activities until the hole in the heart is fixed. Ask a healthcare professional what type and amount of exercise is safest.
* **Extreme altitude changes.** Extreme changes in location above or below sea level may cause complications in people with an unrepaired atrial septal defect. For example, there's less oxygen at higher altitudes. The lower amount of oxygen changes blood flow through the lung arteries. This can cause shortness of breath and strain the heart.
* **Dental work.** If you or your child recently had an ASD fixed and need dental work, talk to a healthcare professional. You or your child may need to take antibiotics for about six months after repair surgery to prevent infection.

**Prevention**

Because the cause of atrial septal defect (ASD) is not clear, prevention may not be possible. But getting good prenatal care is important. If you were born with an ASD, make an appointment for a health checkup before becoming pregnant.

During this visit:

* **Talk about current health conditions and medicines.** It's important to closely control diabetes, lupus and other health conditions during pregnancy. Your healthcare professional may suggest changing doses of some medicines or stopping them before pregnancy.
* **Review your family medical history.** If you have a family history of congenital heart defects or other genetic conditions, you might talk with a genetic counselor to find your risks.
* **Ask about getting tested to see if you've had German measles, also called rubella.** Rubella in a pregnant person has been linked to some types of congenital heart defects in the baby. If you haven't already had German measles or the vaccine, get the recommended vaccinations.

**Prognosis**

If you have an atrial septal defect and are pregnant or thinking about becoming pregnant, talk to a care professional first. It's important to get proper prenatal care. A healthcare professional may suggest repairing the hole in the heart before getting pregnant. A large atrial septal defect or its complications can lead to a high-risk pregnancy.

**complications**

A small atrial septal defect might never cause any concern. Small atrial septal defects often close during infancy.

Larger atrial septal defects can cause serious complications, including:

* Right-sided heart failure.
* Irregular heartbeats, called arrhythmias.
* Stroke.
* Early death.
* High blood pressure in the lung arteries, called pulmonary hypertension.

Pulmonary hypertension can cause permanent lung damage. This complication, called Eisenmenger syndrome, most often occurs over many years. It sometimes happens in people with large atrial septal defects.

Treatment can prevent or help manage many of these complications.

**When to see a doctor**

Serious congenital heart defects are often diagnosed before or soon after a child is born.

Get immediate emergency help if a child has trouble breathing.

Call a healthcare professional if these symptoms occur:

* Shortness of breath, especially during exercise or activity.
* Easy tiring, especially after activity.
* Swelling of the legs, feet or belly area.
* Skipped heartbeats or feelings of a quick, pounding heartbeat.

**Epidemiology**

The prevalence of congenital heart disease, including ASDs, has risen over the past 50 years. Congenital heart disease was diagnosed in less than 1 per 1,000 live births in the 1930s. Recent data indicate a prevalence of 9 per 1,000 live births. Similarly, ASDs were identified in less than 0.5 per 1,000 live births between 1945 and 1949, but recent epidemiologic studies estimate their occurrence at 1.6 per 1,000 live births. This apparent increase in prevalence is likely attributed to advancements in imaging technologies and improved practitioner training rather than a true rise in disease incidence.

Factors associated with the higher prevalence of congenital heart disease include advanced maternal age. Economic and geographical differences also influence diagnosis rates, with congenital heart disease more commonly identified in individuals from developed countries with higher incomes.

**Differential diagnosis**

The differential diagnosis of ASDs includes the following:

* Ventricular septal defect
* Cyanotic congenital heart diseases from sinus venosus and coronary sinus defects
  + Total anomalous pulmonary venous return
  + Pulmonary stenosis
  + Truncus arteriosus
  + Tricuspid atresia

A thorough clinical evaluation and pattern recognition help differentiate ASDs from these conditions, guide treatment decisions, prevent harmful interventions, ensure efficient resource use, and lead to better outcomes.

**REFERENCE**

[**https://www.mayoclinic.org/diseases-conditions/atrial-septal-defect/symptoms-causes/syc-20369715**](https://www.mayoclinic.org/diseases-conditions/atrial-septal-defect/symptoms-causes/syc-20369715)

[**https://www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/atrial-septal-defect-asd**](https://www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/atrial-septal-defect-asd)

[**Atrial Septal Defect - StatPearls - NCBI Bookshelf**](https://www.ncbi.nlm.nih.gov/books/NBK535440/#article-17967.s4)

**Tetralogy of Fallot**

**Definition/Description**

A complex and also rare congenital heart defect that involves four heart malformations (which is to say that, the baby is born with four different heart problems) which lead to an oxygen-poor blood flow out of the heart and into the rest of the body. This simply means that the heart is not formed correctly which leads to the heart pumping blood that is low in oxygen into the rest of the body. These heart problems affect the structure of the heart. The condition causes altered blood flow through the heart and to the rest of the body. Babies with tetralogy of Fallot often have blue or gray skin color due to low oxygen levels

Tetralogy of Fallot is usually diagnosed during pregnancy or soon after the baby is born. If the heart changes and symptoms are mild, tetralogy of Fallot may not be noticed or diagnosed until adulthood.

People who are diagnosed with tetralogy of Fallot need surgery to fix the heart. They will need regular health checkups for life.

**Occurrences:** Tetralogy of Fallot occurs in about 1 out of every 2518 babies born in the U.S. each year.

**Causes**

Tetralogy of Fallot occurs as the baby's heart grows during pregnancy. Usually, the cause is unknown.

Tetralogy of Fallot includes four problems with heart structure:

* Narrowing of the valve between the heart and the lungs, called pulmonary valve stenosis. This condition reduces blood flow from the heart to the lungs. The narrowing may just involve the valve. Or it could happen in more than one place along the pathway between the heart and lungs. Sometimes the valve isn't formed. Instead, a solid sheet of tissue blocks blood flow from the right side of the heart. This is called pulmonary atresia.
* A hole between the bottom heart chambers, called a ventricular septal defect. A ventricular septal defect changes how blood flows through the heart and lungs. Oxygen-poor blood in the lower right chamber mixes with oxygen-rich blood in the lower left chamber. The heart has to work harder to pump blood through the body. The problem may weaken the heart over time.
* Shifting of the body's main artery. The body's main artery is called the aorta. It's usually attached to the left lower heart chamber. In tetralogy of Fallot, the aorta is in the wrong place. It's shifted to the right and sits directly above the hole in the heart wall. This changes how blood flows from the aorta to the lungs.
* Thickening of the right lower chamber of the heart, called right ventricular hypertrophy. When the heart has to work too hard, the wall of the right lower heart chamber gets thick. Over time, this may cause the heart to become weak and eventually fail.

Some people with tetralogy of Fallot have other problems that affect the aorta or heart arteries. There also may be a hole between the heart's upper chambers, called atrial septal defect.

**Risk factors**

The exact cause of tetralogy of Fallot is unknown. Some things may increase the risk of a baby being born with tetralogy of Fallot. Risk factors include:

* Family history.
* Having a virus during pregnancy. This includes rubella, also known as German measles.
* Drinking alcohol during pregnancy.
* Eating poorly during pregnancy.
* Smoking during pregnancy.
* Mother's age is older than 35.
* Down syndrome or DiGeorge syndrome in the baby.

**Signs and symptoms**

Tetralogy of Fallot symptoms depend on how much blood flow is blocked from leaving the heart to go to the lungs. Symptoms may include:

* Blue or gray skin color.
* Shortness of breath and rapid breathing, especially during feeding or exercise.
* Trouble gaining weight.
* Getting tired easily during play or exercise.
* Irritability.
* Crying for long periods of time.
* Fainting.

**Diagnosis methods**

Tetralogy of Fallot is often diagnosed soon after birth. Your baby's skin may look blue or gray. A whooshing sound may be heard when listening to the baby's heart with a stethoscope. This is called a heart murmur.

**Tests**

Tests to diagnose tetralogy of Fallot include:

* **Oxygen level measurement.** A small sensor placed on a finger or toe quickly checks the amount of oxygen in the blood. This is called a pulse oximetry test.
* **Echocardiogram.** This test uses sound waves to create pictures of the heart in motion. It shows the heart and heart valves and how well they are working.
* **Electrocardiogram, also called ECG or EKG.** This test records the electrical activity of the heart. It shows how the heart is beating. Sticky patches called electrodes go on the chest and sometimes the arms or legs. Wires connect the patches to a computer. The computer prints or displays results. An electrocardiogram can help diagnose an irregular heartbeat. Changes in the heart signals also may be due to an enlarged heart.
* **Chest X-ray.** A chest X-ray shows the shape and condition of the heart and lungs. A common sign of tetralogy of Fallot on an X-ray is a boot-shaped heart. That means the right lower chamber is too big.
* **Cardiac catheterization.** This test helps to diagnose or treat certain heart conditions. It may be done to plan surgery. The doctor inserts one or more thin, flexible tubes into a blood vessel, usually in the groin. The tubes are called catheters. The doctor guides the tubes to the heart. During the test, doctors can do different heart tests or treatments.

**Treatment options for Tetralogy of Fallot:**

All babies who have tetralogy of Fallot need surgery to fix the heart and improve blood flow. A heart surgeon, called a cardiovascular surgeon, does the surgery. The timing and type of surgery depends on the baby's overall health and specific heart problems.

Some babies or young children are given medicine while waiting for surgery to keep blood flowing from the heart to the lungs.

**Surgery or other procedures**

Surgery used to treat tetralogy of Fallot may include:

* **Temporary surgery, also called temporary repair.** Some babies with tetralogy of Fallot need a temporary surgery to improve blood flow to the lungs while waiting for open-heart surgery. This type of treatment is called palliative surgery. A surgeon places a tube called a shunt between a large artery that comes off from the aorta and the lung artery. The tube creates a new path for blood to go to the lungs. This surgery may be done if a baby is born early or if the lung arteries aren't fully developed.

The shunt is removed during open-heart surgery to treat tetralogy of Fallot.

* **Open-heart surgery, called complete repair.** People with tetralogy of Fallot need open-heart surgery to completely fix the heart.

A complete repair is usually done in the first year of life. Rarely, a person may not have surgery in childhood if tetralogy of Fallot goes undiagnosed or if surgery is not available. These adults may still benefit from surgery.

A complete repair is done in several steps,

The surgeon patches the hole between the lower heart chambers and repairs or replaces the pulmonary valve. The surgeon may remove thickened muscle below the pulmonary valve or widen the smaller lung arteries.

After complete repair, the right lower chamber won't need to work as hard to pump blood. As a result, the right chamber wall should go back to its usual thickness. The oxygen level in the blood goes up. Symptoms typically get better.

**Lifestyle and home remedies**

After treatment for tetralogy of Fallot, your healthcare team may suggest some steps to keep the heart healthy. These may include:

* **Sports and activity restrictions.** Some people born with a serious heart problem such as tetralogy of Fallot may need to limit exercise or sports activities. But many others can participate in such activities. Ask your or your child's healthcare team which sports and types of activities are safe.
* **Antibiotics to prevent heart infection.** Sometimes, severe heart problems can increase the risk of infection in the lining of the heart or heart valves. This infection is called endocarditis. Antibiotics may be recommended before dental procedures, especially for people who have a mechanical heart valve. Ask your child's healthcare professional if preventive antibiotics are necessary for your child. Good oral care and regular dental checkups also are important ways to help prevent infection.

**Differential Diagnoses**

* Aortic Stenosis
* Bronchiolitis
* Patent Ductus Arteriosus (PDA)
* Pediatric Acute Respiratory Distress Syndrome
* Pediatric Apnea
* Pediatric Foreign Body Ingestion
* Pediatric Pneumonia
* Pneumothorax
* Pulmonic Valvular Stenosis
* Sickle Cell Disease (SCD)

**Prevention tips**

Because the exact cause of most congenital heart defects is unknown, it may not be possible to prevent these conditions. If you have a high risk of giving birth to a child with a congenital heart defect, genetic testing and screening may be done during pregnancy.

There are some steps you can take to help reduce your child's overall risk of birth defects, such as:

* Get proper prenatal care. Regular checkups with a healthcare team during pregnancy can help keep mom and baby healthy.
* Take a multivitamin with folic acid. Taking 400 micrograms of folic acid daily has been shown to reduce birth defects in the brain and spinal cord. It may help reduce the risk of heart defects as well.
* Don't drink or smoke. These lifestyle habits can harm a baby's health. Also avoid secondhand smoke.
* Get a rubella (German measles) vaccine. A rubella infection during pregnancy may affect a baby's heart development. Get vaccinated before trying to get pregnant.
* Control blood sugar. If you have diabetes, good control of your blood sugar can reduce the risk of congenital heart defects.
* Manage chronic health conditions. If you have other health conditions, including phenylketonuria, talk to your healthcare team about the best way to treat and manage them.
* Avoid harmful substances. During pregnancy, have someone else do any painting and cleaning with strong-smelling products.
* Check with your healthcare team before taking any medications. Some medications can cause birth defects. Tell your healthcare team about all the medications you take, including those bought without a prescription.

**Possible Complications**

Untreated tetralogy of Fallot usually leads to life-threatening complications. The complications may cause disability or death by early adulthood.

A possible complication of tetralogy of Fallot is infection of the inner lining of the heart or heart valves. This is called infective endocarditis. Sometimes antibiotics are given before dental work to prevent this type of infection. Ask your healthcare team if preventive antibiotics are right for you or your baby.

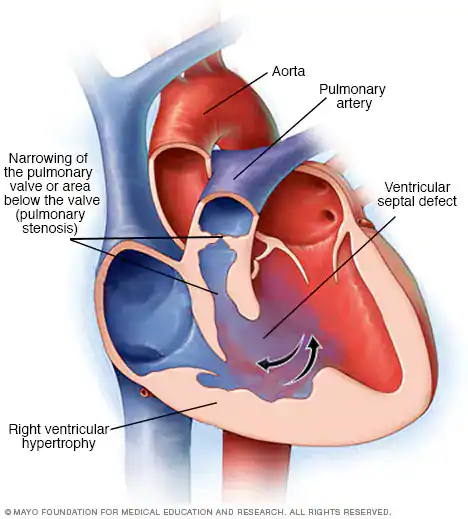
Complications also are possible after surgery to repair tetralogy of Fallot. But most people do well after such surgery. When complications occur, they may include:

* Backward flow of blood through a heart valve.
* Irregular heartbeats.
* A hole in the heart that doesn't go away after surgery.
* Changes in the size of the heart chambers.
* Swelling of part of the aorta, called aortic root dilation.
* Sudden cardiac death.

Another procedure or surgery may be needed to fix these complications.

Congenital heart defects and pregnancy

People born with a complex congenital heart defect may be at risk for complications during pregnancy. Talk to your healthcare team about the possible risks and complications of pregnancy. Together you can discuss and plan for any special care needed.



**When to see a doctor**

Serious congenital heart defects are often diagnosed before or soon after your child is born. Seek medical help if you notice that your baby has these symptoms:

* Trouble breathing.
* Bluish color of the skin.
* Lack of alertness.
* Seizures.
* Weakness.
* More irritable than usual.

If your baby becomes blue or gray, place your baby on the side and pull the baby's knees up to the chest. This helps increase blood flow to the lungs. Call 911 or your local emergency number immediately.

**Epidemiology**

Tetralogy of Fallot (TOF) represents approximately 7-10% of congenital heart diseases (CHDs),and it is the most common cyanotic CHD, with 0.23-0.63 cases per 1000 births.This disorder accounts for one third of all CHD in patients younger than 15 years; in adults, tetralogy of Fallot has an estimated prevalence of 1 in 3500 to 1 in 4300 people.

In most cases, tetralogy of Fallot is sporadic and nonfamilial. The incidence in siblings of affected parents is 1-5%, and it occurs more commonly in males than in females. The disorder is associated with extracardiac anomalies such as cleft lip and palate, hypospadias, and skeletal and craniofacial abnormalities. Genetic studies indicate that in some patients with tetralogy of Fallot, there may be 22q11.2 deletion and other submicroscopic copy number alterations.

Adult patients with tetralogy of Fallot currently represent a very large group of patients who underwent congenital heart surgery in early life. Although the exact number of these adults is not known, because many are lost to follow-up or have never been followed, it is estimated that over two thirds of affected children who undergo repair of tetralogy of Fallot in early childhood will reach adulthood. One study showed a 94% survival rate of 168 patients aged 16 years and older who underwent simple repair.

Tetralogy of Fallot (TOF) is one of the most common congenital heart diseases in Nigeria, accounting for **approximately 11.8% of all congenital heart diseases among children in the country**. A study conducted in a tertiary hospital in South Western Nigeria between 2007 and 2014 found that TOF is as prevalent in Nigeria as in other parts of the world.

In Nigeria, TOF is often diagnosed later in life due to limited access to diagnostic facilities and expertise. Cyanosis is the most common presenting feature and indication for evaluation. The male to female ratio for TOF in Nigeria is approximately 1.7.3

The prevalence of TOF among congenital heart diseases in Nigeria varies, with some studies reporting rates of 10%-26.2%. In a study from an urban center in Africa, the prevalence of TOF among congenital heart diseases was found to be 15.8%.

It is important to note that Nigeria, like other low- and middle-income countries, faces challenges in diagnosing and treating TOF due to limited resources, financial means, and expertise. This often results in patients undergoing surgery much later than in high-income countries, leading to complications such as right ventricular dysfunction, cardiac arrhythmias, and poor psychomotor development.

There is a need to establish more cardiac centers in Nigeria to improve early diagnosis and better outcomes for children with TOF. Collaboration from developed countries can be helpful in addressing these resource limitations.

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**Reference**

John Hopkins Medicine by Pediatric cardiologist Rukmalee Vithana

<https://emedicine.medscape.com/article/2035949-differential?form=fpf>

<https://www.hopkinsmedicine.org/health/conditions-and-diseases/tetralogy-of-fallot-tof>

<https://emedicine.medscape.com/article/2035949-overview#a7>

**Patent Ductus Arteriosus (PDA)**

**Definition and description**

Patent ductus arteriosus (PDA) is a persistent opening between the two major blood vessels leading from the heart.

An opening called the ductus arteriosus is part of the blood flow system in the womb for an unborn baby, also called a fetus. It usually closes shortly after birth. If the opening remains open, it's called a patent ductus arteriosus. A PDA is a type of congenital heart defect.

A small PDA might never need treatment. But a large, untreated patent ductus arteriosus can let oxygen-poor blood move the wrong way. This can make the heart muscle weak, causing heart failure and other complications.

**Causes**

The exact causes of patent ductus arteriosus and other congenital heart defects are not clear. During the first six weeks of pregnancy, an unborn baby's heart starts to form and beat. The major blood vessels to and from the heart grow. It's during this time that certain congenital heart defects may begin to develop.

Before birth, there is a temporary opening called the ductus arteriosus between the two main blood vessels leaving a baby's heart. Those vessels are the aorta and the pulmonary artery. The opening is needed for a baby's blood flow before birth. It moves blood away from a baby's lungs while they develop. The baby gets oxygen from the mother's blood.

After birth, the ductus arteriosus is no longer needed. It usually closes within 2 to 3 days. But in some infants, the opening doesn't close. When it stays open, it's called a patent ductus arteriosus.

The constant opening causes too much blood to flow to the baby's lungs and heart. Untreated, the blood pressure in the baby's lungs might increase. The baby's heart might grow larger and get weak.

**Risk factors**

Risk factors for patent ductus arteriosus (PDA) include:

* Premature birth: Patent ductus arteriosus occurs more commonly in babies who are born too early than in babies who are born full term.
* Family history and other genetic conditions: A family history of heart conditions present at birth may increase the risk of PDA. Babies born with an extra chromosome 21, a condition called Down syndrome, also are more likely to have patent ductus arteriosus.
* German measles during pregnancy: Having German measles, also called rubella, during pregnancy can cause changes in an unborn baby's heart. A blood test done before pregnancy can tell if you're immune to rubella. A vaccine is available for those who aren't immune.
* Being born at a high altitude: Babies born above 8,200 feet (2,499 meters) have a greater risk of PDA than babies born at lower altitudes.
* Female sex assigned at birth: Patent ductus arteriosus is twice as common in girls.

**Signs and symptoms**

Patent ductus arteriosus symptoms (PDA) depend on the size of the opening and the person's age. A small PDA might not cause symptoms. Some people don't notice symptoms until adulthood. A large PDA can cause symptoms of heart failure soon after birth.

A large PDA found during infancy or childhood might cause:

* Poor eating, which leads to poor growth.
* Sweating with crying or eating.
* Constant fast breathing or being out of breath.
* Easy tiring.
* Rapid heart rate

**Diagnosis methods**

To diagnose patent ductus arteriosus (PDA), a healthcare professional does a physical exam and asks questions about you or your child's medical history. The healthcare professional may hear a heart sound called a murmur while listening to the heart with a stethoscope.

**Tests**

Tests can help diagnose patent ductus arteriosus.

* **Echocardiogram.** Sound waves make pictures of the beating heart. This test shows how blood flows through the heart and heart valves. It can tell if there is a persistent opening in the heart. An echocardiogram also can tell if there are higher pressures in the lung arteries.
* **Chest X-ray.** This test shows the condition of the heart and lungs.
* **Electrocardiogram (ECG or EKG).** This quick and simple test records the electrical signals that make up the heartbeat. It shows how fast or how slow the heart is beating.
* **Cardiac catheterization.** This test isn't usually needed to diagnose patent ductus arteriosus. But it might be done if there also are other heart conditions. A doctor places a thin, flexible tube called a catheter into a blood vessel, usually in the groin or wrist, and guides it to the heart. During this test, treatments may be done to close the patent ductus arteriosus.

**Treatment options**

Treatments for patent ductus arteriosus (PDA) depend on the age of the person being treated.

Some people with small PDAs may only need regular health checkups to watch for complications. A premature baby born with patent ductus arteriosus also needs regular checkups to make sure the opening closes.

**Medications**

A premature baby born with patent ductus arteriosus (PDA) may get medicines called nonsteroidal anti-inflammatory drugs. These medicines, also called NSAIDs, block certain body chemicals that keep the PDA open. However, these medicines won't close a PDA in full-term babies, children or adults.

In the past, healthcare professionals told people born with patent ductus arteriosus to take antibiotics to prevent infection before dental work and some surgeries. This is no longer recommended for most people with patent ductus arteriosus. Ask your healthcare professional whether preventive antibiotics are necessary. They might be recommended after certain heart procedures.

**Surgery or other procedures**

Treatments to close a patent ductus arteriosus include:

* **Using a thin tube called a catheter** and a plug or coil to close the opening. This treatment is called a catheter procedure. It lets a doctor repair the opening without open-heart surgery.

To do this treatment, the doctor places the catheter into a blood vessel in the groin and guides it to the heart. A plug or coil goes through the catheter. The plug or coil closes the patent ductus arteriosus. The treatment doesn't usually need an overnight hospital stay.

Premature babies are too small for catheter treatments. If the PDA isn't causing trouble, a catheter treatment to close the opening may be done when the baby is older.

* **Open-heart surgery** to close the PDA. This treatment is called surgical closure. It may be needed if medicine doesn't work or if the PDA is large or causing complications.

A surgeon makes a small cut between the ribs to reach the heart. The PDA is closed using stitches or clips. It usually takes a few weeks to fully recover from this surgery.

Some people born with PDA need regular health checkups for life, even after treatment to close the opening. During these checkups, a healthcare professional may do tests to check for complications. Talk with your healthcare professional about your care plan. Ideally, it's best to seek care from a doctor trained in treating adults with heart conditions before birth. This type of doctor is called a congenital cardiologist.

**Prevention tips**

There is no known prevention for patent ductus arteriosus. However, it's important to do everything possible to have a healthy pregnancy. Here are some of the basics:

* **Get early prenatal care, even before you're pregnant.** If you're thinking about pregnancy, talk with your healthcare team to develop a prenatal care plan. Also tell your healthcare professional about all the medicines you take, including those bought without a prescription.
* **Start taking folic acid.** Taking 400 micrograms of folic acid daily before and during pregnancy has been shown to reduce brain and spinal cord conditions in the baby. It also may help reduce the risk of heart conditions.
* **Exercise and stay active.** Work with your healthcare professional to develop an exercise plan that's right for you.
* **Do not drink alcohol or smoke.** These lifestyle habits can harm a baby's health. Also avoid secondhand smoke.
* **Get recommended vaccines.** Update your vaccinations before becoming pregnant. Certain types of infections can be harmful to a developing baby.
* **Control blood sugar.** If you have diabetes, good control of your blood sugar may reduce the risk of certain heart conditions present at birth in your baby.

**Lifestyle and home remedies**

Anyone born with a patent ductus arteriosus needs to take steps to keep the heart healthy and prevent complications. These tips can help.

* **Don't smoke.** Smoking is a major risk factor for heart disease and other heart conditions. Quitting is the best way to reduce the risk. If you need help quitting, talk with your healthcare professional.
* **Eat healthy foods.** Eat plenty of fruits, vegetables and whole grains. Limit sugar, salt and saturated fats.
* **Practice good hygiene.** Regularly wash your hands and brush and floss your teeth to keep yourself healthy.
* **Ask about sports limits.** Some people born with heart conditions may be told not to do some types of exercise or sports activities. Ask your healthcare professional which sports and types of exercise are safe for you or your child.
* **Manage stress.** Find ways to help reduce emotional stress. Some tips are to get more exercise, practice mindfulness and connect with others in support groups. If you have anxiety or depression, talk with your healthcare professional about treatments that can help.

**EPIDEMIOLOGY**

The commonest congenital heart diseases in Nigeria are ventricular septal defect (40.6%), patent ductus arteriosus (18.4%), atrial septal defect (11.3%) and tetralogy of Fallot (11.8%). There has been a 6% increase in the burden of VSD in every decade for the 5 decades studied and a decline in the occurrence of pulmonary stenosis. Studies conducted in Northern Nigeria demonstrated higher proportions of atrial septal defects than patent ductus arteriosus.

Ventricular septal defects are the commonest congenital heart diseases in Nigeria with a rising burden.

In Nigeria, the epidemiology of Patent Ductus Arteriosus (PDA) shows that it is one of the more common congenital heart defects, following ventricular septal defect (VSD) and atrial septal defect (ASD) in frequency. According to studies conducted between 1964 and 2015, PDA accounts for approximately 18.4% of congenital heart diseases in children in Nigeria.

The prevalence of PDA in Nigeria has been studied across different regions and time periods. Ventricular septal defects (VSD) have shown a rising trend over the past five decades, while there has been a decline in the occurrence of pulmonary stenosis. In Northern Nigeria, studies have demonstrated higher proportions of atrial septal defects than PDA.

The incidence of PDA is higher in preterm infants and those with respiratory distress syndrome. In extremely premature infants, up to 80% may have a PDA at 3 days of age.

Prognosis

Optimal treatment remains contentious. Options include conservative/medical, pharmacologic, and surgical management. Conservative/medical management includes mild fluid restriction, increased airway pressures, and supportive care. Pharmacologic treatment is accomplished using indomethacin, ibuprofen, or acetaminophen. Surgical intervention is by direct closure or by percutaneous ligation. Treatment may be prophylactic, presymptomatic, or symptomatic. Long-term morbidities associated with PDA include chronic lung disease, retinopathy of prematurity, and neurodevelopmental delay.

Absence of a universal scoring system for severity of PDA limits accuracy of comparisons among research studies. Lack of a consistent definition also makes it difficult to aggregate data for meta-analyses. Adoption of a consistent scoring system for hemodynamic significance would facilitate comparisons of outcomes among research studies.

Clinicians should be aware of treatment options for PDA and their implications on neonatal outcomes. For nurses, anticipation of possible side effects is important for performance of focused assessments.

With treatment, most babies born with PDA live healthy and active lives.

If PDA doesn’t close on its own, healthcare providers can correct it, if needed. Babies and children with moderate and large sized PDA that are not treated in the correct time frame may be at higher risk for developing heart complications as adults. Talk with your healthcare provider about whether your baby needs follow-up care.

**Possible complications**

A small patent ductus arteriosus might not cause complications. Larger, untreated PDAs could cause:

* **High blood pressure in the lungs, also called pulmonary hypertension.** A large PDA causes irregular blood flow in the heart and lungs. As a result, pressure goes up in the pulmonary artery. Over time, the increased pressure damages the smaller blood vessels in the lungs. A life-threatening and lasting type of lung damage called Eisenmenger syndrome may occur.
* **Heart failure.** Symptoms of this serious complication include rapid breathing, often with gasping breaths, and poor weight gain.
* **Heart infection, called endocarditis.** Patent ductus arteriosus can increase the risk of germs attaching to areas in the heart. Without quick treatment, endocarditis can damage or destroy the heart valves.

**Patent ductus arteriosus and pregnancy**

It may be possible to have a successful pregnancy with a small patent ductus arteriosus. However, having a large PDA or a complication such as heart failure, lung damage or irregular heartbeats increases the risk of serious complications during pregnancy.

Before becoming pregnant, talk with your healthcare professional about possible pregnancy risks and complications. Some heart medicines can be dangerous for an unborn baby. Your healthcare professional may stop or change your medicines before you become pregnant.

Together you can plan for any special care needed during pregnancy. If you are at high risk of having a baby with a heart condition present at birth, genetic testing and screening may be done during pregnancy.

**When to see a doctor / red flag**

Contact a healthcare professional if your baby or older child:

* Tires easily when eating or playing.
* Isn't gaining weight.
* Becomes breathless when eating or crying.
* Always breathes fast or is short of breath.

**Differential diagnosis (how it’s distinguished from other illnesses)**

Other conditions that should be considered when evaluating a patient with suspected patent ductus arteriosus (PDA) include the following:

* Absence pulmonary valve syndrome
* Acute anemia
* Aortic regurgitation
* Aortopulmonary window (aortopulmonary fenestration)
* Atrioventricular malformation
* Bacteremia and sepsis
* Bronchial pulmonary artery stenosis
* Cardiogenic shock
* Cervical venous hum (usually present on the right side of the neck and more prominent in the sitting position, varying with respiration)
* Dilated cardiomyopathy
* Mitral regurgitation
* Ruptured sinus of Valsalva and fistula
* Peripheral pulmonary artery stenosis
* Persistent truncus arteriosus
* Pulmonary arteriovenous fistula
* Systemic arteriovenous fistula (cerebrovascular or hepatic arteriovenous malformations)
* Total anomalous pulmonary venous return
* Venous hum
* Ventricular septal defect (VSD) with aortic regurgitation

REFERENCES

<https://pubmed.ncbi.nlm.nih.gov/27605952/>

<https://my.clevelandclinic.org/health/diseases/17325-patent-ductus-arteriosus-pda>

<https://emedicine.medscape.com/article/891096-differential?form=fpf>

**Hypertensive heart disease**

**Heart problems caused by high blood pressure**

High blood pressure, also known as hypertension, can lead to several heart problems over time. These include heart attack, stroke, and heart failure.

Heart attacks can occur when the arteries that supply blood to the heart become blocked due to high blood pressure, which can cause the arteries to harden and narrow.24 High blood pressure can also cause the heart to work harder, leading to thickening of the heart muscle, a condition known as left ventricular hypertrophy. If this condition progresses, the heart may not get enough oxygen, causing angina (chest pain), and eventually, heart failure.

Heart failure is a condition where the heart cannot pump enough blood to meet the body's needs. High blood pressure makes the heart work harder, causing it to get bigger and struggle to pump enough blood to the body. Symptoms of heart failure include shortness of breath, fatigue, swelling in the ankles, lower legs, or abdomen, and difficulty sleeping when lying flat.

In addition to these, high blood pressure can also cause an aneurysm, which is a bulge in a weakened blood vessel. If an aneurysm ruptures, it can be life-threatening.

It's important to manage high blood pressure to prevent these complications. Treatment usually aims to control symptoms and slow down the progression of the condition.

**Definition and description**

It is also called Hypertension-hypertensive heart, High blood pressure-hypertensive heart. It refers to the problems that occur because of high blood pressure that is present over a long time.

Blood pressure is the force of the blood pushing against the artery walls. The force is generated with each heartbeat as blood is pumped from the heart into the blood vessels. The size and elasticity of the artery walls also affect blood pressure. Each time the heart beats (contracts and relaxes), pressure is created inside the arteries.

The pressure is greatest when blood is pumped out of the heart into the arteries. When the heart relaxes between beats (blood is not moving out of the heart), the pressure falls in the arteries.

Two numbers are recorded when measuring blood pressure.

* The top number, or **systolic pressure**, refers to the pressure inside the artery when the heart contracts and pumps blood through the body.
* The bottom number, or **diastolic pressure**, refers to the pressure inside the artery when the heart is at rest and is filling with blood.

Both the systolic and diastolic pressures are recorded as "mm Hg" (millimeters of mercury). This recording represents how high the mercury column in the blood pressure cuff is raised by the pressure of the blood.

Blood pressure is measured with a blood pressure cuff and stethoscope by a nurse or other healthcare provider. You can also take your own blood pressure with an electronic blood pressure monitor. These are available at most pharmacies.

The National Heart, Lung, and Blood Institute (NHLBI) of the National Institutes of Health (NIH) has determined 2 levels of high blood pressure for adults:

* **Stage 1**
  + 140 mm Hg to 159 mm Hg systolic pressure—higher number

and

* + 90 mm Hg to 99 mm Hg diastolic pressure—lower number
* **Stage 2**
  + 160 mm Hg or higher systolic pressure

and

* 100 mm Hg or higher diastolic pressure

The NHLBI defines prehypertension as:

* 120 mm Hg to 139 mm Hg systolic pressure

and

* 80 mm Hg to 89 mm Hg diastolic pressure

The NHLBI guidelines define normal blood pressure as follows:

* Less than 120 mm Hg systolic pressure

and

* Less than 80 mm Hg diastolic pressure
* The American College of Cardiology and the American Heart Association divide blood pressure into four general categories. Ideal blood pressure is categorized as normal.
* **Normal blood pressure.** Blood pressure is lower than 120/80 mm Hg.
* **Elevated blood pressure.** The top number ranges from 120 to 129 mm Hg and the bottom number are below, not above, 80 mm Hg.
* **Stage 1 hypertension.** The top number ranges from 130 to 139 mm Hg or the bottom number is between 80- and 89-mm Hg.
* **Stage 2 hypertension.** The top number is 140 mm Hg or higher or the bottom number is 90 mm Hg or higher.
* Blood pressure higher than 180/120 mm Hg is considered a hypertensive emergency or crisis. Seek emergency medical help for anyone with these blood pressure numbers.
* Untreated, high blood pressure increases the risk of heart attack, stroke and other serious health problems. It's important to have your blood pressure checked at least every two years starting at age 18. Some people need more-frequent checks.
* Healthy lifestyle habits —such as not smoking, exercising and eating well — can help prevent and treat high blood pressure. Some people need medicine to treat high blood pressure.

Use these numbers as a guide only. A single elevated blood pressure measurement is not necessarily an indication of a problem. Your healthcare provider will want to see multiple blood pressure measurements over several days or weeks before making a diagnosis of high blood pressure and starting treatment. If you normally run a lower-than-usual blood pressure, you may be diagnosed with high blood pressure with blood pressure measurements lower than 140/90.

**What Is Hypertensive Heart Disease?**

Your heart beats around 100,000 times every day. Every minute, it pumps nearly 1.5 gallons of blood. But there are a lot of reasons why this crucial organ can stop working properly. One of those is chronic hypertension, or what you might know as ongoing high blood pressure.

“Hypertensive heart disease is a condition where the heart muscle can get damaged over time due to long-standing high blood pressure,” says Joy Gelbman, MD, a cardiologist at Weill Cornell Medicine and New York-Presbyterian. “The heart can become big, thickened or weakened due to pumping against a high blood pressure over a long time.”

**Causes**

About 1 out of 3 Americans have high blood pressure (higher than 120/80). Only half of these cases are managed well.

You won’t be able to feel if blood is pushing against the walls of your arteries with too much force. That’s why high blood pressure is sometimes called a “silent killer.” But over time, hypertension can make it harder for your heart to pump enough blood. It can also damage or narrow your blood vessels. That can become an even bigger issue if you also have high cholesterol. Cholesterol is a fatty substance that can clog the pathways through which blood flows in your body.

Usually, chronic hypertension happens because of several different factors. These can include:

* Your genes. If at least one of your close family members has high blood pressure, you may be at a higher risk.
* Tobacco use. This includes smoking, vaping, and smokeless tobacco.
* Food choices. For instance, a diet high in sodium can put extra pressure on your heart.
* Lack of exercise
* Heavy alcohol use
* Drug use. Recreational drugs, such as cocaine and amphetamines, can raise your blood pressure and keep it high.
* Certain medications. For example, some drugs that affect your immune system can cause what’s called “secondary hypertension.”
* Other ongoing health conditions. These include sleep apnea and kidney disease.

High blood pressure that isn’t treated for a long time can lead to hypertensive heart disease (HTN heart disease.) You could be at a higher risk if:

* You’re over the age of 45.
* You’re overweight.
* You don’t get enough exercise.
* You have diabetes.
* You have high cholesterol.
* You drink alcohol.
* You eat a lot of salt.
* You use tobacco.

Hypertension puts extra strain on your heart, which may cause it to weaken and become stiff. Over time, it can begin to fail.

If your arteries become so damaged that enough blood can’t reach your heart, you could have a condition called ischemic heart disease (IHD).

**Some other causes of hypertensive heart disease are**:

* High blood pressure means the pressure inside the blood vessels (called arteries) is too high. As the heart pumps against this pressure, it must work harder. Over time, this causes the heart muscle to thicken.
* Because there are often no symptoms with high blood pressure, people can have the problem without knowing it. Symptoms most often do not occur until after many years of poor blood pressure control, when damage to the heart has occurred.
* Eventually, the muscle may become so thick that it does not get enough oxygen. This can cause angina (chest pain). Without appropriate blood pressure control, the heart can weaken over time and heart failure may develop.
* High blood pressure also leads to thickening of the blood vessel walls. When combined with cholesterol deposits in the blood vessels, the risk of heart attack and stroke increases.
* Hypertensive heart disease is the leading cause of illness and death from high blood pressure.

Blood pressure is determined by two things: the amount of blood the heart pumps and how hard it is for the blood to move through the arteries. The more blood the heart pumps and the narrower the arteries, the higher the blood pressure.

There are two main types of high blood pressure.

Primary hypertension, also called essential hypertension

For most adults, there's no identifiable cause of high blood pressure. This type of high blood pressure is called primary hypertension or essential hypertension. It tends to develop gradually over many years. Plaque buildup in the arteries, called atherosclerosis, increases the risk of high blood pressure.

Secondary hypertension

This type of high blood pressure is caused by an underlying condition. It tends to appear suddenly and cause higher blood pressure than does primary hypertension. Conditions and medicines that can lead to secondary hypertension include:

* Adrenal gland tumors
* Blood vessel problems present at birth, also called congenital heart defects
* Cough and cold medicines, some pain relievers, birth control pills, and other prescription drugs
* Illegal drugs, such as cocaine and amphetamines
* Kidney disease
* Obstructive sleep apnea
* Thyroid problems

Sometimes just getting a health checkup causes blood pressure to increase. This is called white coat hypertension.

**Risk factors**

Nearly one-third of all Americans have high blood pressure, but it is particularly prevalent in:

* People who have diabetes, gout, or kidney disease
* African Americans (particularly those who live in the southeastern U.S.)
* People in their early to middle adult years; men in this age group have higher blood pressure more often than women in this age group
* People in their middle to later adult years; women in this age group have higher blood pressure more often than men in this age group (more women have high blood pressure after menopause than men of the same age)
* Middle-aged and elderly people; more than half of all Americans age 60 and older have high blood pressure
* People with a family history of high blood pressure
* People consuming a high salt diet
* Women who are taking oral contraceptives
* People with depression
* High blood pressure has many risk factors, including:
* **Age.** The risk of high blood pressure increases with age. Until about age 64, high blood pressure is more common in men. Women are more likely to develop high blood pressure after age 65.
* **Race.** High blood pressure is particularly common among Black people. It develops at an earlier age in Black people than it does in white people.
* **Family history.** You're more likely to develop high blood pressure if you have a parent or sibling with the condition.
* **Obesity or being overweight.** Excess weight causes changes in the blood vessels, the kidneys and other parts of the body. These changes often increase blood pressure. Being overweight or having obesity also raises the risk of heart disease and its risk factors, such as high cholesterol.
* **Lack of exercise.** Not exercising can cause weight gain. Increased weight raises the risk of high blood pressure. People who are inactive also tend to have higher heart rates.
* **Tobacco usage or vaping.** Smoking, chewing tobacco or vaping immediately raises blood pressure for a short while. Tobacco smoking injures blood vessel walls and speeds up the process of hardening of the arteries. If you smoke, ask your care provider for strategies to help you quit.
* **Too much salt.** A lot of salt — also called sodium — in the body can cause the body to retain fluid. This increases blood pressure.
* **Low potassium levels.** Potassium helps balance the amount of salt in the body's cells. A proper balance of potassium is important for good heart health. Low potassium levels may be due to a lack of potassium in the diet or certain health conditions, including dehydration.
* **Drinking too much alcohol.** Alcohol use has been linked with increased blood pressure, particularly in men.
* **Stress.** High levels of stress can lead to a temporary increase in blood pressure. Stress-related habits such as eating more, using tobacco or drinking alcohol can lead to further increases in blood pressure.
* **Certain chronic conditions.** Kidney disease, diabetes and sleep apnea are some of the conditions that can lead to high blood pressure.
* **Pregnancy.** Sometimes pregnancy causes high blood pressure.
* High blood pressure is most common in adults. But kids can have high blood pressure too. High blood pressure in children may be caused by problems with the kidneys or heart. But for a growing number of kids, high blood pressure is due to lifestyle habits such as an unhealthy diet and lack of exercise.

**Signs and symptoms**

Most people with high blood pressure have no symptoms, even if blood pressure readings reach dangerously high levels. You might not have any symptoms for years, until some damage has been done to your heart. The signs of hypertensive cardiovascular disease include:

* Chest pain
* Headaches
* Nosebleed
* Feeling short of breath
* Feeling dizzy
* Fainting
* Heart palpitations (fast, pounding, or fluttering heartbeat)
* Fatigue (feeling tired for no reason)
* Trouble doing regular activities
* Having a hard time breathing (especially when lying down)
* Swelling in your lower legs

However, these symptoms aren’t specific. They usually don’t occur until high blood pressure has reached a severe or life-threatening stage.

**Diagnosis methods**

Your doctor will ask about your family history of heart issues, and check your blood pressure. If you’re on medication for high blood pressure, they’ll likely ask if you’re taking your medication as prescribed and if you are checking your blood pressure at home. If so, your doctor will want to know some of your recent readings.

They’ll also ask about any symptoms you’re having. During a physical exam, your doctor can listen to your heart. They’ll likely also order some tests, including:

* Blood work and urine (pee) tests to get a better idea of your overall health
* Electrocardiogram (EKG), a quick in-office test that checks the electrical activity of your heart
* Echocardiogram, an imaging test that can show any changes in the structure of your heart
* MRI (Magnetic Resonance Imaging), another noninvasive test that uses strong magnetic fields magnetic field gradients, and radio waves to form images of the organs in the body; images that provide a detailed view of soft tissues, such as the brain or abdomen, and can show organs, bones, muscles, and blood vessels; these images can offer many specific details about how your heart is working

**Treatment options**

“When you’re diagnosed with hypertensive heart disease, there are still measures you can take to prevent further complications,” says Carmen W. Landrau, MD, a cardiologist and spokesperson for the American Heart Association.

The first step in treating hypertensive cardiovascular disease is treating the high blood pressure that’s causing it. That can be done in several ways, including prescribed medications and making small changes to your daily habits.

Medications for hypertensive heart disease

Many different treatments can help manage your blood pressure. They work by relaxing or widening your blood vessels, helping to flush excess fluid from your body, or slowing down your heart rate so your heart doesn’t have to work as hard. Your doctor could prescribe:

* Diuretics
* Beta-blockers
* Angiotensin-converting enzyme (ACE) inhibitors
* Calcium channel blockers
* Angiotensin receptor blockers (ARBs)
* Vasodilators
* Renin inhibitors

**Lifestyle changes for hypertensive heart disease**

Your doctor may also advise you to make changes to your lifestyle. You can:

Make healthy food choices. That includes limiting the amount of sodium and saturated fats you eat. Fill your plate instead with more fruits, vegetables, whole grains, and low-fat dairy.

More physical activity. Working out helps control your blood pressure, maintains your heart function, and helps you reach and stay at a healthy weight.

Avoid tobacco products and alcohol. If you need help quitting, ask your doctor for tips.

Get regular checkups. During follow-up visits, your doctor can make sure you and your heart are staying healthy.

Manage your stress level. Feeling anxious can raise your blood pressure.

**Surgeries for hypertensive heart disease**

If your condition doesn’t get better with medication and lifestyle changes, your doctor may talk to you about a medical procedure.

For instance, an FDA-approved procedure called renal ablation, or renal denervation, helps some people who have resistant hypertension.

During this minimally invasive surgery, a cardiologist uses ultrasound or radiofrequency energy to damage the renal nerves in your kidneys. Making these nerves less active can help control your blood pressure.

Other types of surgeries can help strengthen your arteries or repair or replace damaged parts of your heart. Your doctor can explain which kind of surgery is right for you.

**Prevention tips**

To try to prevent hypertensive cardiovascular disease, you can:

**Have regular checkups.** Your blood pressure should be checked by your doctor at least once a year, more often if it’s high.

**Take care of any other health conditions.** For instance, manage your blood sugar if you live with diabetes. If you know your cholesterol is high, make changes to your eating habits to try to bring it down.

**Enjoy healthy meals.** Try to eat fewer processed foods, as they’re often high in sodium. Instead, look for fresh, flavorful recipes that fit into the Mediterranean or DASH (Dietary Approaches to Stop Hypertension) diets.

**Get moving.** When you work out regularly, you can lower your blood pressure by 5 to 8 points. Aim for about 30 minutes of physical activity every day, but even less than that is good for you. Every bit counts.

**Quit tobacco and curb your alcohol intake.** Doing so could not only lower your risk of hypertensive heart disease but also improve your overall health.

**Don’t skimp on sleep.** Sleeping less than seven hours a night regularly can raise your blood pressure. If you have trouble sleeping, talk to your doctor.

**Keep stress in check.** You can’t get rid of all the stress in your life, but you can get better at dealing with it. If you need new ways to manage stress, a counselor or therapist can help.

**Prognosis**

What can I expect if I have hypertensive heart disease?

Hypertensive heart disease is a long-term disease that takes years to develop. Over time, people who have it are at a higher and higher risk of dying from a cardiovascular problem. The prognosis for people with hypertensive cardiovascular disease is different from person to person, depending on

* What symptoms you’re having.
* Whether you have cardiovascular disease or risk factors.
* Other medical conditions you have.

**Possible complications**

Hypertensive Heart Disease Complications

Getting and keeping your blood pressure under control can reduce your chances of more severe heart issues.

Heart failure

Heart failure, or congestive heart failure, doesn’t mean your heart stops working. It means that your heart’s pumping power is less than it should be. With heart failure, blood doesn’t move well through your heart’s chambers, and pressure in your heart increases, making it harder for your heart to deliver oxygen and nutrients to your body.

At first, the chambers of your heart will stretch to try to keep blood moving. Your body will also try to adjust in other ways. For instance, your blood vessels will narrow to increase the force of blood flow. And your kidneys will retain more fluid (water) and sodium so there’s a larger amount of blood in your body. But these are only short-term fixes.

Symptoms of heart failure include:

* Shortness of breath
* Fatigue
* Finding it hard to do everyday tasks (such as carrying groceries)
* Excess coughing
* Wheezing
* Swelling in your legs, feet, fingers, or belly
* Not feeling hungry/lack of appetite
* Increased heart rate
* Nausea (feeling sick to your stomach)
* Fatigue

**Ischemic heart disease (IHD)**

* When your arteries can no longer open wide enough to easily let blood through, your heart becomes damaged. One of the most common symptoms of ischemic heart disease is chest pain, or what’s called angina.

**Coronary artery disease (CAD)**

* This is another name for ischemic heart disease. You could also hear it called coronary heart disease.

**Left ventricle hypertrophy (LVH)**

* High blood pressure makes your heart work harder than it should, which causes its walls to thicken. As a result, the left ventricle (pumping chamber) starts to get stiff and weak. That can prevent enough blood flow, although you may not have symptoms for many years.

**Enlarged heart**

* Your doctor will be able to see this on an imaging test. An enlarged heart isn’t a condition. But it is a sign that your heart is working harder than normal or has been damaged.

**Stroke**

* A stroke happens when blood flow to your brain gets blocked. It’s a medical emergency because your brain cells can start dying in minutes. Stroke symptoms include having trouble seeing, speaking, walking, or understanding others, numbness on one side of your body, or a sudden, severe headache. If you think you’re having a stroke, call 911 right away. Don’t wait to see if your symptoms get better or go away on their own.

**Sudden cardiac arrest (SCA)**

* If your heart stops working because of an abnormal heart rhythm, you’ll collapse and stop breathing. SCA is different from a heart attack, which happens when blood stops flowing to your heart. CPR can help get your heart started again. An electrical device called an automated external defibrillator (AED) can also shock your heart into pumping again.

Some other complications of Hypertensive heart disease include:

The excessive pressure on the artery walls caused by high blood pressure can damage blood vessels and body organs. The higher the blood pressure and the longer it goes uncontrolled, the greater the damage.

Uncontrolled high blood pressure can lead to complications including:

* **Heart attack or stroke.** Hardening and thickening of the arteries due to high blood pressure or other factors can lead to a heart attack, stroke or other complications.
* **Aneurysm.** Increased blood pressure can cause a blood vessel to weaken and bulge, forming an aneurysm. If an aneurysm ruptures, it can be life-threatening.
* **Heart failure.** When you have high blood pressure, the heart has to work harder to pump blood. The strain causes the walls of the heart's pumping chamber to thicken. This condition is called left ventricular hypertrophy. Eventually, the heart can't pump enough blood to meet the body's needs, causing heart failure.
* **Kidney problems.** High blood pressure can cause the blood vessels in the kidneys to become narrow or weak. This can lead to kidney damage.
* **Eye problems.** Increased blood pressure can cause thickened, narrowed or torn blood vessels in the eyes. This can result in vision loss.
* **Metabolic syndrome.** This syndrome is a group of disorders of the body's metabolism. It involves the irregular breakdown of sugar, also called glucose. The syndrome includes increased waist size, high triglycerides, decreased high-density lipoprotein (HDL or "good") cholesterol, high blood pressure and high blood sugar levels. These conditions make you more likely to develop diabetes, heart disease and stroke.
* **Changes with memory or understanding.** Uncontrolled high blood pressure may affect the ability to think, remember and learn.
* **Dementia.** Narrowed or blocked arteries can limit blood flow to the brain. This can cause a certain type of dementia called vascular dementia. A stroke that interrupts blood flow to the brain also can cause vascular dementia.

**When to see a doctor / red flag**

* Blood pressure screening is an important part of general health care. How often you should get your blood pressure checked depends on your age and overall health.
* Ask your provider for a blood pressure reading at least every two years starting at age 18. If you're 40 or older, or you're 18 to 39 with a high risk of high blood pressure, ask for a blood pressure check every year.
* Your care provider will likely recommend more-frequent readings if you have high blood pressure or other risk factors for heart disease.
* Children age 3 and older may have blood pressure measured as a part of their yearly checkups.
* If you don't regularly see a care provider, you may be able to get a free blood pressure screening at a health resource fair or other locations in your community. Free blood pressure machines are also available in some stores and pharmacies. The accuracy of these machines depends on several things, such as a correct cuff size and proper use of the machines. Ask your health care provider for advice on using public blood pressure machines.

Contact your health care provider if you have high blood pressure and develop any symptoms

**Differential diagnosis (how it’s distinguished from other illnesses)**

Several conditions can mimic the clinical, ECG, or echocardiographic features of HHD, making accurate diagnosis essential. Differentiating these entities, which include the conditions below, is critical to avoid misclassification and ensure appropriate management.

* Severe aortic stenosis
* Hypertrophic cardiomyopathy
* Infiltrative cardiomyopathies (eg, amyloidosis, sarcoidosis)
* Ischemic heart disease, including HFpEF or systolic dysfunction
* Athlete’s heart
* Sleep apnea
* Restrictive cardiomyopathies characterized by LAE and diastolic dysfunction

Recognizing these mimickers requires careful integration of clinical context, imaging findings, and, when necessary, advanced diagnostic testing. Early identification of alternative diagnoses can significantly influence prognosis and treatment strategy.

**Outlook / Prognosis**

What can I expect if I have hypertensive heart disease?

Hypertensive heart disease is a long-term disease that takes years to develop. Over time, people who have it are at a higher and higher risk of dying from a cardiovascular problem. The prognosis for people with hypertensive cardiovascular disease is different from person to person, depending on:

* What symptoms you’re having.
* Whether you have cardiovascular disease or risk factors.
* Other medical conditions you have.

Living With

How do I take care of myself with hypertensive heart disease?

When you have high blood pressure, it’s important to keep taking your medicines to regulate your blood pressure. You also need to watch for problems that start to develop and treat them promptly.

Conditions to watch for include:

* Ventricular hypertrophy.
* Atrial fibrillation.
* Angina.
* Heart failure.

What questions should I ask my doctor?

* What is my personal risk of hypertensive heart disease?
* Is there anything else I can do to reduce my risk of hypertensive heart disease?

**EPIDEMIOLOGY**

There were 190 diagnoses in the 163 patients with 27 patients having a double diagnosis, consisting of 88 (54%) males and 75 (46%) females. The mean age was 50.4 years (age range 9-85 years). Ten types of acquired heart pathologies were identified and they included hypertensive heart disease in 49.47%, rheumatic heart disease in 26.32%, cardiomyopathy in 11.05%, endomyocardial fibrosis in 4.74%, and pericarditis in 3.68%. Others were cor pulmonale, pulmonary hypertension, intracardiac thrombi, left atrial myxoma and degenerative heart disease which accounted for the remaining 4.74%.

The odds ratios (OR) for myocardial infarction, overall stroke, ischaemic stroke, and intracerebral hemorrhagic stroke associated with hypertension in both men and women were included 15,152 cases with acute myocardial infarction and 14,820 controls from across 262 centers in 52 regions worldwide; a total of 578 cases and 789 controls were from Africa.

663 and 2237 cases of intracerebral hemorrhagic stroke and ischaemic stroke, respectively, and 3,000 controls from 84 centers in 22 countries. A total of 141 and 214 cases of intracerebral hemorrhagic stroke and ischemic stroke were respectively reported for Africa (Nigeria, South Africa, Sudan, and Uganda were among the African countries included). A self-reported history of hypertension was used to define hypertension in both studies.

**Statistical Analysis**

The population attributable fraction (PAF%) was estimated using the Levin formula shown below

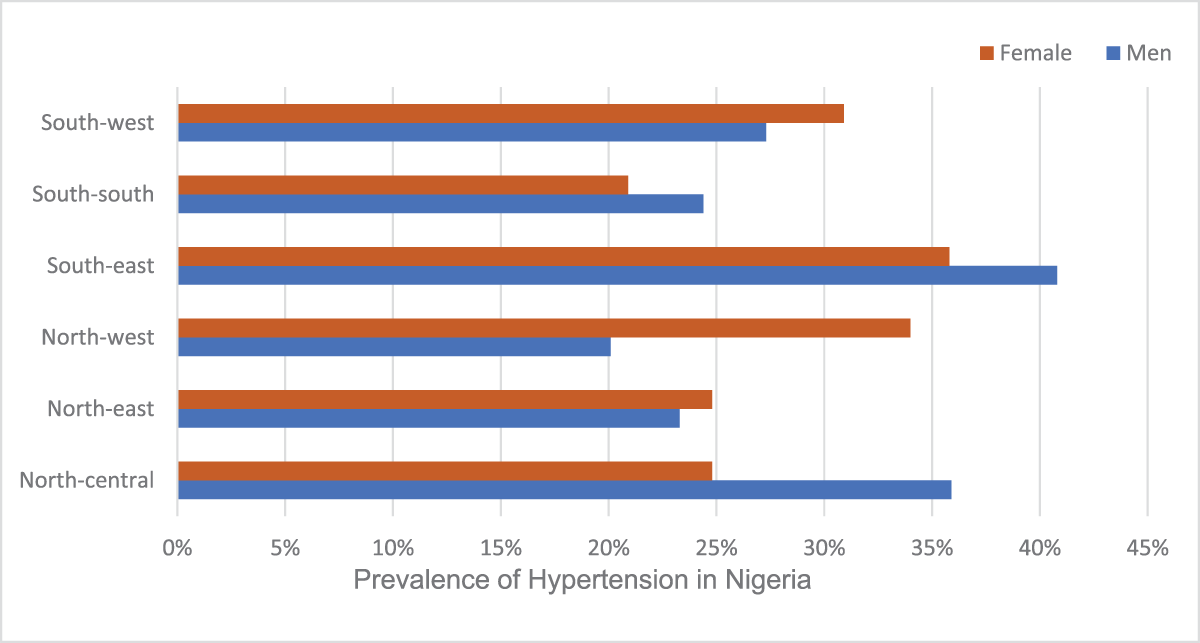
PAF(%)=Pe (RR−1)[Pe (RR−1)+1]×100%

Where; Pe is the probability of exposure to the risk factor, that is, the prevalence of hypertension in Nigeria and RR is the relative risk. Estimation of relative risks from the ORs was done using the equation below, and subsequently, the PAF for the burden of CVDs in Nigeria was obtained.

RR=OR(1−Pe)+(Pe∗OR)

We estimated the above PAF% for the burden of CVDs overall and across subgroups defined by age, sex, and geopolitical zone. Sex-specific PAFs% were calculated for all the outcomes. For myocardial infarction, age- and region-specific PAFs% were computed based on the information available. PAF% for all the outcomes for both 1995 and 2020 were also computed. A student t-test was used to evaluate the significance of the differences in PAF across the region. The test was conducted at a two-sided 5% level of significance with no adjustment for multiple testing. Statistical analysis was carried out using R version 4.1.0 (2021-05-18).

The reported prevalence of hypertension in Nigeria was 30.9% and ranged across the regions from 24.7% (North West) to 33.3% (South East). Men in the South East had the highest pooled prevalence of hypertension (40.8%) compared to women (35.1%), and this was also among women in the South East. The lowest prevalence of hypertension was in the North West for men (20.1%) and in the South South for women (20.9%) .

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Prevalence of hypertension in the six geopolitical zones in Nigeria

Sex-specific PAFs for all outcomes

Table 1 shows the sex-specific PAF of myocardial infarction, all stroke, ischaemic stroke and intracerebral hemorrhagic stroke in the six geopolitical zones of Nigeria. Overall, 13.2% of the burden of myocardial infarction and 24.6% of the burden of all strokes could be attributed to hypertension, with slight variation between the regions. Hypertension was estimated to have contributed about 21.6% and 33.1% of the burden of ischaemic stroke and intracerebral hemorrhagic stroke, respectively.

Table 1

Sex-Specific PAF (%) of myocardial infarction, all stroke, ischaemic stroke and intracerebral hemorrhagic stroke in the six geopolitical zones in Nigeria.

## **References**

Bakris GL, Sorrentino MJ. Systemic hypertension: mechanisms, diagnosis, and treatment. In: Libby P, Bonow RO, Mann DL, Tomaselli GF, Bhatt DL, Solomon SD, eds.*Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine*. 12th ed. Philadelphia, PA: Elsevier; 2022:chap 26.

Rogers JG, O'Connor CM. Heart failure: pathophysiology and diagnosis. In: Goldman L, Schafer AI, eds. *Goldman-Cecil Medicine*. 26th ed. Philadelphia, PA: Elsevier; 2020:chap 52.

Siu AL, US Preventive Services Task Force. Screening for high blood pressure in adults: US Preventive Services Task Force recommendation statement.*Ann Intern Med*. 2015;163(10):778-786. PMID: 26458123 [**pubmed.ncbi.nlm.nih.gov/26458123/**](https://pubmed.ncbi.nlm.nih.gov/26458123/).

Victor RG. Arterial hypertension. In: Goldman L, Schafer AI, eds. *Goldman-Cecil Medicine*. 26th ed. Philadelphia, PA: Elsevier; 2020: chap 70.

Whelton PK, Carey RM, Aronow WS, et al. 2017 ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA guideline for the prevention, detection, evaluation, and management of high blood pressure in adults: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2018;71(19):e127-e248. PMID: 29146535 [**pubmed.ncbi.nlm.nih.gov/29146535/**](https://pubmed.ncbi.nlm.nih.gov/29146535/).

<http://my.clevelandclinic.org/health/diseases/21840-hypertensive-heart-disease>

<https://my.clevelandclinic.org/health/diseases/21840-hypertensive-heart-disease#outlook-prognosis>

[Hypertensive Heart Disease - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK539800/#article-23225.s9)

**Hypertensive Crisis (Urgency/Emergency)**

**Definition and description**   
Hypertensive crises are an umbrella term for hypertensive urgency and hypertensive emergency. These two occur when blood pressure becomes very high, possibly causing organ damage.

During a hypertensive crisis, the heart may not be able to pump blood as well as it should.

There are two types of hypertensive crises.

* Urgent hypertensive crisis. Blood pressure is 180/120 mm Hg or greater. There are no signs of organ damage.
* Emergency hypertensive crisis. Blood pressure is 180/120 mm Hg or greater. There is life-threatening damage to the body's organs.

**Hypertensive Urgency**

Hypertensive urgency occurs when blood pressure spikes -- blood pressure readings are 180/110 or higher -- but there is no damage to the body's organs or symptoms. Blood pressure can be brought down safely within a few hours with blood pressure medication.

**Hypertensive Emergency**

Hypertensive emergency means blood pressure is so high that organ damage can occur. Blood pressure must be reduced immediately to prevent imminent organ damage.

Organ damage associated with hypertensive emergency may include:

* Changes in mental status, such as confusion
* Bleeding into the brain (stroke)
* Heart failure
* Chest pain (unstable angina)
* Fluid in the lungs (pulmonary edema)
* Heart attack
* Aneurysm (aortic dissection)
* Eclampsia (occurs during pregnancy)

Hypertensive emergencies are rare. When it does occur, it is often when hypertension goes untreated, if the patient does not take their blood pressure medication, or they have taken an over-the-counter medication that exacerbates high blood pressure.

**Causes**

Causes of a hypertensive crisis include:

* Forgetting to take blood pressure medicine.
* Suddenly stopping certain heart medicines, including beta blockers.
* Medicine interactions.
* Tumor of the adrenal gland, called a pheochromocytoma.

**Risk factors**

Age and sex impact the risk of the onset of a hypertensive emergency. Older adults face higher odds of experiencing hypertensive emergencies compared to younger people.

The reasons include age-related changes that affect blood pressure regulation and a higher likelihood of having comorbidities, two or more diseases simultaneously. Men also have increased susceptibility compared to women, possibly due to hormonal influences.

If you are an older male, you are in the highest-risk demographic category for a hypertensive emergency. Therefore, it is important to monitor and treat your blood pressure carefully. Work closely with your doctor to stabilize your blood pressure, particularly if you have other risk factors.

You may have a higher risk of having a hypertensive crisis if you:

* Have obesity
* Are male
* Are Black
* Don’t take your blood pressure medicines consistently
* Use stimulant drugs that aren’t prescribed for you

**Signs and symptoms**

Symptoms of a hypertensive emergency may include:

* Headache or blurred vision
* Anxiety
* Increasing confusion
* Nausea and vomiting
* Not responding to stimulation
* Seizure
* Increasing chest pain
* Increasing shortness of breath
* Swelling or edema (fluid buildup in the tissues)

If you get a very high blood pressure reading at home and don't have any symptoms, relax for a few minutes. Then check your blood pressure again. If it's still very high, seek medical care.

Call emergency medical services if your blood pressure is 180/120 mm Hg or greater and you have chest pain, shortness of breath, or symptoms of stroke. Stroke symptoms include:

* Numbness or tingling.
* Loss of feeling in the face, arm or leg, often on just one side of the body.
* Trouble walking.
* Trouble speaking.
* Changes in vision.

Treatment for a hypertensive crisis may include a hospital stay to watch for organ damage. Medicines to lower blood pressure are given by mouth or IV.

**Diagnosis methods**

To diagnose a hypertensive emergency, your health care providers will ask you several questions to get a better understanding of your medical history, such as whether you're taking blood pressure medication or have recently eaten something salty. They will also need to know all medications you are taking, including nonprescription and recreational drugs. Also, be sure to tell them if you are taking any herbal or dietary supplements.

Certain tests will be performed to monitor blood pressure and assess organ damage, including:

* Regular monitoring of blood pressure
* Eye exam to look for swelling and bleeding
* Blood and urine testing

**How doctors diagnose this condition**

A healthcare provider will take your blood pressure in both arms to diagnose a hypertensive crisis. They’ll also review your medical history, talk to you about any symptoms you have and do a physical exam. You may need some tests to help your provider find the cause.

Possible tests include:

* Blood tests
* Urine tests
* Electrocardiogram (EKG/ECG)
* Fundoscopic exam
* Neurological exam
* Transthoracic echocardiogram
* Computed tomography (CT) scan of your chest and head

Your provider will diagnose you with a hypertensive emergency if you have signs of new or worsening organ damage. They’ll diagnose you with hypertensive urgency if these signs aren’t there.

**Treatment**

In a hypertensive emergency, the first goal is to bring down the blood pressure as quickly as possible with intravenous (IV) blood pressure medications to prevent further organ damage. Whatever organ damage has occurred is treated with therapies specific to the organ that is damaged.

**How is a hypertensive crisis treated?**

Treatment for a hypertensive crisis happens in the emergency room. Healthcare providers give you medicine to bring your blood pressure down to a safe level.

You may go home the same day (with medicines to take on your own) if you don’t have signs of organ damage. But if you’re having a hypertensive emergency, you’ll need to stay in the intensive care unit (ICU) for a couple of days. Providers will give you medicine directly into your veins (through an IV). They’ll also continuously monitor your blood pressure.

Your care team will decide how quickly to bring down your blood pressure based on what other medical conditions you have. In some cases, lowering your blood pressure too quickly can prevent your organs and tissues from getting enough blood. So, your providers may lower it gradually over 24 to 48 hours.

But they’ll bring down your blood pressure more quickly during a hypertensive crisis if you have certain conditions like aortic dissection, severe preeclampsia or eclampsia. In these cases, the benefits of rapid lowering outweigh any risks.

**Medications**

Medicines for hypertensive crisis treatment include:

* Captopril
* Clevidipine
* Clonidine
* Esmolol
* Hydralazine
* Labetalol
* Nicardipine
* Nifedipine
* Nitroglycerin
* Nitroprusside

Providers choose the right medicine for your needs. They take into account your medical conditions and other medicines you’re taking.

Prevention tips

Practice healthy living habits to help prevent high blood pressure

Eat a healthy diet

Choose healthy meal and snack options to help you avoid high blood pressure and its complications. Be sure to eat plenty of fresh fruits and vegetables.

Talk with your health care team about eating a variety of foods rich in potassium, fiber, and protein and lower in salt (sodium) and saturated fat. For many people, making these healthy changes can help keep blood pressure low and protect against heart disease.

Keep yourself at a healthy weight

Being overweight or obese increases your risk for high blood pressure.

Talk with your health care team about ways to reach a healthy weight, including choosing healthy foods and getting regular physical activity.

Be physically active

Physical activity can help keep you at a healthy weight and lower your blood pressure. The Physical Activity Guidelines for Americans recommends that adults get at least 2 hours and 30 minutes of moderate-intensity exercise, such as brisk walking or bicycling, every week. That's about 30 minutes a day, 5 days a week. Children and adolescents should get 1 hour of physical activity every day.

Do not smoke

Smoking raises your blood pressure and puts you at higher risk for heart attack and stroke. If you do not smoke, do not start. If you do smoke, quitting will lower your risk for heart disease. Your doctor can suggest ways to help you quit.

Limit how much alcohol you drink

Do not drink too much alcohol, which can raise your blood pressure. Men should have no more than 2 alcoholic drinks per day, and women should have no more than 1 alcoholic drink per day.

Get enough sleep

Getting enough sleep is important to your overall health. It also helps keep your heart and blood vessels healthy. Not getting enough sleep on a regular basis is linked to an increased risk of heart disease, high blood pressure, and stroke.

Manage stress

People who have depression, anxiety, stress, or post-traumatic stress disorder over a long period of time may develop other health problems, including an increased heart rate and high blood pressure.

Recognize the signs and symptoms of mental health disorders and heart disease. Talk with your health care team about potential heart conditions in relation to your mental health.

**Prognosis**

A hypertensive crisis is a warning sign that your blood pressure isn’t where it needs to be. Your provider will help you lower your blood pressure and keep it at healthy levels. Make sure to keep all your follow-up appointments and take your medicines exactly as your provider prescribes them

**Complications of a hypertensive crisis**

Dangerously high blood pressure can lead to:

* A sudden, rapid decline in heart function (acute heart failure)
* Sudden fluid buildup in your lungs (acute pulmonary edema)
* Sudden loss of kidney function (acute kidney failure)
* A tear in your largest artery (aortic dissection)
* Bleeding around your brain (intracranial hemorrhage)
* Lack of blood flow to your heart (heart attack)
* Lack of blood flow to your brain (ischemic stroke)
* Temporary brain dysfunction (hypertensive encephalopathy)

**When to see a doctor / red flag**

Hypertension symptoms? Often there aren't any. High blood pressure is often called the "silent" disease, because it may have no noticeable symptoms.

If undetected and untreated, hypertension can cause heart disease (including congestive heart failure and heart attack), stroke, and kidney disease. That's why it is important to have regular physical examinations to make sure your blood pressure is within the normal range. This is especially important if your blood pressure has ever been high, if you have a family history of hypertension, or if you are gaining weight.

If you are being treated for high blood pressure, your doctor can answer any questions or concerns you may have during your regular visits. However, there may be situations that warrant a call to your doctor. For example:

* If you aren't responding to the treatment your doctor prescribed, and your blood pressure is still high
* If you have certain symptoms, including fatigue, nausea, shortness of breath, lightheadedness, headache, excessive sweating, palpitations or irregular heartbeats, problems with your vision, or confusion, these may be serious and should warrant prompt medical attention. They could be from uncontrolled high blood pressure or from medication side effects.

If you have any concerns about your condition, don't hesitate to call your doctor.

**Differential diagnosis (how it’s distinguished from other illnesses)**

The following symptoms and differential diagnoses are examples of what your healthcare provider might consider before making a final diagnosis. Contact your healthcare provider if you experience symptoms to diagnose and treat your condition.

**Abdominal pain**

Symptoms of abdominal pain include an ache, cramps or sharp pains at mild to severe levels localized to a specific area in your stomach region. A differential diagnosis of abdominal pain includes:

* Appendicitis.
* Gastritis.
* Inflammatory bowel disease.
* Intestinal or bowel blockage.
* Pancreatitis.

**Asthma**

Symptoms of asthma include shortness of breath, wheezing, pain or a tight feeling in your chest and coughing. A differential diagnosis of asthma includes:

* Allergic rhinitis.
* Bronchitis.
* Chronic obstructive pulmonary disease (COPD).
* Pneumonia.

**Back pain**

Symptoms of back pain include aching, burning or sharp, stabbing pain that increases when standing, walking, lifting objects or twisting. A differential diagnosis of back pain includes:

* Arthritis.
* Disk hernia.
* Fibromyalgia.
* Osteoporosis.

**Chest pain**

Symptoms of chest pain include aching, sharp pain, burning sensations, tightness or squeezing pressure localized in your chest area. A differential diagnosis of chest pain includes:

* Angina.
* Anxiety.
* Muscle strain.
* Pneumonia.
* Viral infection.

**Cough**

Symptoms of a cough include clearing your throat of mucus or fluids and clearing your throat of dry air, irritation or tickle in the back of your throat. A differential diagnosis of cough includes:

* Asthma.
* Bronchitis.
* Pneumonia.
* Reflux.
* Seasonal allergies.

**Depression**

Symptoms of depression include fatigue, low energy, anxiety, showing a range of emotions and appetite changes. A differential diagnosis of depression includes:

* Anxiety.
* Bipolar disorder.
* Dementia.
* Hypothyroidism.

**Elevated alkaline phosphatase**

Symptoms of elevated alkaline phosphatase include stomach or abdominal pain, nausea and vomiting and a yellow color to the skin (jaundice). A differential diagnosis of elevated alkaline phosphatase includes but is not limited to:

* Blockage in the liver, gall bladder or bile ducts.
* Gallstones.
* Liver disease.

**Fatigue**

Symptoms of fatigue include feeling tired, lack of energy, not getting enough sleep and feeling weak throughout your entire body. A differential diagnosis of fatigue includes but is not limited to:

* Anemia.
* Depression.
* Insomnia.
* Thyroid disease.

**Headache**

Symptoms of a headache include head pain that ranges in severity from mild to severe, a throbbing sensation and sensitivity to lights and sounds with symptoms that last for hours or up to several days. A differential diagnosis of a headache includes:

**Hypertension**

Symptoms of hypertension include chest pain, headaches, dizziness, shortness of breath and feeling tired. A differential diagnosis of hypertension includes:

* Kidney disease.
* Sleep apnea.
* Thyroid disease.

**Knee pain**

Symptoms of knee pain include swelling around the knee, instability when standing, stiffness and popping noises when you move your knee. A differential diagnosis of knee pain includes:

* Arthritis.
* Cartilage tear.
* Osteoarthritis.
* Strained ligaments.
* Tendonitis.

**Urinary tract infection (UTI)**

Symptoms of a urinary tract infection include feeling like you need to pee even when you have an empty bladder, feeling a burning sensation when you pee and urinating often. A differential diagnosis of a urinary tract infection includes:

* Chlamydia.
* Gonorrhea.
* Interstitial cystitis.
* Vaginal yeast infection.

**Epidemiology data (e.g., how common it is, affected demographics).**

38% of adult Nigerians aged 18 years and above are hypertensive. Out of the hypertensive subjects, 60% were aware of their status, one-third were receiving treatment and 12% had their blood pressure under control. Prevalence of hypertension ranged from 20.9% in the North-Central to 52.8% in the South-East region. Hypertension is as common in the rural as in the urban areas; however, the urban dwellers are more aware of and receive treatment for the condition more than their rural counterparts.

The survey included 19,997 Nigerians drawn from about 13 states of the country and spread across rural and urban communities. The investigators used a threshold blood pressure of 160/95 mmHg for the diagnosis of hypertension and reported that the prevalence of hypertension was 11.2%, 14% and 9.8% in overall, urban and rural areas respectively. In terms of the geographic spread, the semi-desert area corresponding to the North-West geopolitical region had the highest prevalence of hypertension.

The narrowing urban-rural gap in the prevalence of hypertension may be related to the increasing urbanization of Nigerian rural areas with the attendant shift towards the lifestyle that fuels non-communicable diseases including hypertension. Population growth results in dwindling land for farming thus encouraging rural dwellers to engage in more non-farm economic activities]. The non-farm activities as opposed to the manual labor driven farm activities encourage sedentary lifestyle and consumption of high-salt processed food. A review of previous studies that evaluated the urban-rural difference in the prevalence of hypertension in South-East Nigeria, elsewhere in sub-Saharan Africa; Southern America, and China suggest either a narrowing of the rural urban disparity or even a trend towards a higher prevalence in the rural compared to the urban areas in some of the studies. Although prevalence of hypertension is obviously increasing in the rural populations of Nigeria, the rate of awareness and treatment has remained significantly higher in the urban areas as compared to the rural areas. This is consistent across the aforementioned studies conducted in different regions of the world.

hypertension in Nigeria is among the highest in Africa. Several factors ranging from access to health care, poor health seeking behaviour, dysfunctional health system, therapeutic inertia and patients’ non-adherence to prescribed therapies may be responsible for the huge loss in care of hypertension.

**References:**

1. Hypertensive crisis: When you should call 911 for high blood pressure. American Heart Association. https://www.heart.org/en/health-topics/high-blood-pressure/understanding-blood-pressure-readings/hypertensive-crisis-when-you-should-call-911-for-high-blood-pressure. Accessed June 13, 2022.
2. Ferri FF. Hypertension. In: Ferri's Clinical Advisor 2023. Elsevier; 2023. https://www.clinicalkey.com. Accessed July 15, 2022.
3. Elliott WJ. Evaluation and treatment of hypertensive emergencies in adults. https://www.uptodate.com/contents/search. Accessed June 14, 2022.
4. Varon J, et al. Management of severe asymptomatic hypertension (hypertensive urgencies) in adults. https://www.uptodate.com/contents/search. Accessed June 14, 2022.
5. Whelton PK, et al. 2017 ACC/AHA/AAPA/ABC/ACPM/AGS/APhA/ASH/ASPC/NMA/PCNA guideline for the prevention, detection, evaluation, and management of high blood pressure in adults: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Hypertension. 2018; doi:10.1161/HYP.0000000000000065.
6. Lopez-Jimenez F (expert opinion). Mayo Clinic. July 6, 2022.
7. Benenson I, et al. Risk factors for hypertensive crisis in adult patients: A systematic review. JBI Evidence Synthesis. 2021; doi:10.11124/JBIES-20-00243.
8. <https://icfamilymedicine.com/hypertensive-emergency-risk-factors/>
9. <https://my.clevelandclinic.org/health/diseases/24470-hypertensive-crisis>
10. [Differential Diagnosis: Definition and Examples](https://my.clevelandclinic.org/health/diagnostics/22327-differential-diagnosis)

**Pulmonary heart disease (cor pulmonale)**

**Right heart failure due to lung disease**

* It is also known as Right ventricular hypertrophy, or cor pulmonale.

**Definition and description**

Cor pulmonale is a loosely defined term that describes the change in structure or function of the right ventricle of the heart. The right ventricle of the heart is responsible for carrying deoxygenated blood to the lungs.

A primary underlying lung condition causes cor pulmonale. It typically presents itself in chronic, slowly progressive ways but can also be onset and acute. Issues with the left side of the heart are never considered cor pulmonale.

Cor pulmonale is an enlarged right ventricle in your heart that happens because of a lung condition. Pushing against high pressure in your pulmonary artery can cause your right ventricle to fail. Treatment addresses the lung condition that caused cor pulmonale. Most cases are chronic, or long-term.

This is when a lung issue causes your right ventricle (heart chamber) to get so big that your heart starts to fail. It’s a type of right-sided heart failure

Normally, your right ventricle sends blood to your pulmonary artery to get oxygen from your lungs. But certain lung conditions cause high blood pressure in your pulmonary artery. That makes it harder for your right ventricle to get blood to your lungs.

Your right ventricle has to push against higher pressure in your pulmonary artery, where blood pressure is normally low. It’s like opening a car door when it’s windy outside. You have to push against the wind.

When your right ventricle has to keep working harder than normal, it gets larger and doesn’t work well.

**Types of cor pulmonale**

Types of cor pulmonale include:

* Acute cor pulmonale (short-term).
* Chronic cor pulmonale (long-term).

**Causes**

Cor pulmonale causes include lung conditions that cause pulmonary hypertension, such as:

* Chronic obstructive pulmonary disease (COPD). This is the top cause of long-term cor pulmonale.
* Pulmonary embolism (blood clot).
* Trauma or surgery that removes lung tissue.
* Mechanical ventilation injury.
* Scleroderma.
* Cystic fibrosis.
* Neuromuscular disorders that affect breathing.
* Lung issues that block your veins.
* Pulmonary fibrosis (scarring).
* Obstructive sleep apnea.
* Kyphosis (curved spine).
* Primary pulmonary hypertension

**Risk factors**

Risk factors for cor pulmonale include:

* Using tobacco products.
* Breathing polluted air.
* Having lung conditions that cause pulmonary hypertension.

**Signs and symptoms**

Some symptoms of cor pulmonale might include:

* Cyanosis: A change of the body tissue color to a bluish-purple hue, as a result of decrease in the amount of oxygen bound to the hemoglobin in the red blood cells of the capillary bed.
* Wheezing

Cor pulmonale symptoms may also include:

* Shortness of breath.
* Bulging veins in your neck.
* Swelling in your legs or belly.
* Fatigue.
* Chest pain.
* Fainting episodes.

At first, you may not have cor pulmonale symptoms. They happen when you’ve had cor pulmonale for a while. Symptoms of cor pulmonale occur in addition to the symptoms of the lung condition that caused it.

**Diagnosis methods**

A healthcare provider may hear abnormal sounds when they listen to your heart with a stethoscope. They’ll order tests to diagnose cor pulmonale.

What tests will be done to diagnose cor pulmonale?

Tests to diagnose cor pulmonale may include:

* Echocardiogram.
* Chest X-ray.
* Electrocardiogram (EKG).
* Nuclear medicine imaging.
* CT angiogram.
* Heart MRI.
* VQ scan.
* Right heart catheterization.
* Thrombophilia screen

**Treatment option**

Cor pulmonale treatment focuses on improving the lung issue that caused it.

The goal is to:

* Get more oxygen to your tissues if they need it.
* Help your right ventricle work better.
* Keep the blood vessels in your lungs from constricting too much so blood can get through more easily.

Quick treatment is important to prevent permanent damage to your heart.

Specific medicines or procedures used

Cor pulmonale treatments include:

* Oxygen therapy.
* Bronchodilators.
* Corticosteroids.
* Diuretics.
* Anticoagulants.
* Cardiac rehab.
* Blood clot removal (embolectomy).
* Pulmonary vasodilators.
* Lung or heart-lung transplant.

Complications/side effects of the treatment

Complications or side effects of treatments may include:

| Treatment | Side effect or complication |
| --- | --- |
| Oxygen therapy | Headaches; Fatigue; Nosebleeds. |
| Medicines | Upset stomach; Weight gain; Mood changes; Headache; Bleeding. |
| Cardiac rehab | Injury; Abnormal heart rhythm. |
| Embolectomy | Bleeding; Injury; Infection. |
| Lung transplant | Bleeding; Infection; Organ rejection. |

* Vasodilators
* Diuretics

**Prevention tips for Cor pulmonale**

Can cor pulmonale be prevented?

You can prevent cor pulmonale by not getting the lung conditions that cause it. You can lower your risk of cor pulmonale by:

Avoiding tobacco products and smoke.

Avoiding air pollution.

Avoiding things that cause pulmonary hypertension.

Getting prompt treatment for lung conditions.

**Prognosis of Cor Pulmonale**

**What can I expect if I have cor pulmonale?**

Most people with cor pulmonale have a poor quality of life. Even with treatment, symptoms often come back.

How long cor pulmonale lasts

Cor pulmonale is usually a long-term condition. However, cor pulmonale from a mechanical ventilation injury or pulmonary embolism is a short-term condition.

**Outlook for cor pulmonale**

The condition that causes cor pulmonale determines the outlook for people who have it. Without management of the lung condition that caused it, the outlook is poor. For instance, only 30% of people who get cor pulmonale from COPD live another five years.

**Possible complications**

Complications of cor pulmonale may include:

* Abnormal heart rhythms.
* Right heart failure.
* Low oxygen levels (hypoxia).
* Fainting episodes.
* Kidney disease.
* Liver disease.
* Death.

**When to see a doctor / red flag**

If you spent time in the hospital for cor pulmonale, you’ll need to follow up with a cardiologist and a pulmonologist (heart and lung specialists). They may want to see you in a week or two.

**Differential diagnosis (how it’s distinguished from other illnesses)**

* Atrial myxoma
* Blood disorders that are associated with increased blood viscosity
* Chronic thromboembolic pulmonary hypertension
* Congestive (biventricular) heart failure
* Constrictive pericarditis
* High-output heart failure
* Infiltrative cardiomyopathies
* Interstitial lung disease (ILD)
* Obstructive sleep apnea (OSA)
* Primary pulmonic stenosis
* Pulmonary hypertension
* Right heart failure due to congenital heart diseases
* Right heart failure due to right ventricular infarction
* Ventricular septal defect

**Epidemiology data**

The exact prevalence of cor pulmonale is difficult to determine, as physical examination and routine tests are relatively insensitive for the detection of pulmonary hypertension and RV dysfunction. Cor pulmonale is estimated to account for 6% to 7% percent of all types of adult heart disease in the United States. The incidence of cor pulmonale is widely variant among countries. It depends on air pollution, the prevalence of cigarette smoking and other risk factors for various lung diseases.

Reference

<https://my.clevelandclinic.org/health/diseases/24922-cor-pulmonale>

<https://www.ncbi.nlm.nih.gov/books/NBK430739/#article-19975.s9>

**Heart murmurs**

**Alternative Names**

Chest sounds – murmurs

Heart sounds – abnormal

Murmur – innocent

Innocent murmur

Systolic heart murmur

Diastolic heart murmur

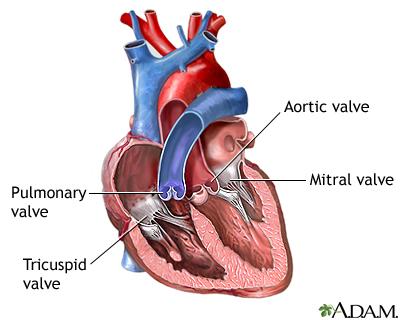
Heart murmurs are sounds — such as whooshing or swishing — made by rapid, choppy (turbulent) blood flow through the heart. The sounds can be heard with a device called a stethoscope. A typical heartbeat makes two sounds like "lubb-dupp" (sometimes described as "lub-DUP") when the heart valves are closing. Heart murmurs can be present at birth (congenital) or develop later in life (acquired).

Some heart murmurs are harmless (innocent). An innocent heart murmur is not a sign of heart disease and doesn't need treatment. Other heart murmurs may be a sign of a serious heart condition. Tests are needed to check the heart and heart valves. Heart murmur treatment depends on the cause.

Description

Murmurs can happen for many reasons, such as:

* When a valve does not close tightly and blood leaks backward (regurgitation)
* When blood flows through a narrowed or stiff heart valve (stenosis)



There are several ways in which your health care provider may describe a murmur:

* Murmurs are classified ("graded") depending on how loud the murmur sounds with a stethoscope. The grading is on a scale. Grade I can barely be heard and is intermittent. An example of a murmur description is a "grade II/VI murmur." (This means the murmur is grade 2 on a scale of 1 to 6).

**Heart murmur grading**

Doctors grade heart murmurs by how intense or loud they are. The systolic grading system is for murmurs that happen when your heart squeezes. It ranges from 1 to 6:

* Grade 1 is so faint that your doctor can barely hear it with a stethoscope in a quiet room.
* Grade 2 is faint, but your doctor can hear it with a stethoscope.
* Grade 3 is loud enough for your doctor to hear easily with a stethoscope.
* Grade 4 is very loud and obvious.
* Grade 5 is so loud that your doctor can hear it with only one side of the stethoscope touching your chest.
* Grade 6 is so loud that your doctor can hear it when the stethoscope isn't touching your chest.

The diastolic grading system is for murmurs that happen when your heart relaxes. It ranges from 1 to 4:

* Grade 1 is so quiet your doctor can barely hear it.
* Grade 2 is soft, but your doctor can hear it.
* Grade 3 is easy to hear.
* Grade 4 is loud and clear.
* In addition, a murmur is described by the stage of the heartbeat when the murmur is heard. A heart murmur may be described as systolic or diastolic. (Systole is when the heart is squeezing out blood and diastole is when it is filling up with blood.)

When a murmur is more noticeable, the provider may be able to feel it with the palm of the hand over the heart. This is called a "thrill" and means the murmur is grade 4 or higher.

**What Are the Different Types of Murmurs?**

Your child's health care provider will evaluate a murmur based on several factors. Murmurs are analyzed for pitch, loudness, and duration. They also are graded according to their intensity (on a scale of one to six, with one being very faint and six being very loud).

Types of murmurs include the following:

* **Systolic murmur.**A heart murmur that occurs during a heart muscle contraction. Systolic murmurs are divided into ejection murmurs (often due to blood flow through a narrowed vessel or irregular valve) and regurgitant murmurs (typically due to mitral or tricuspid regurgitation where the blood leaks back into the atria from the ventricles).
* **Diastolic murmur.**A heart murmur that occurs during heart muscle relaxation between beats. Diastolic murmurs are due to a narrowing (stenosis) of the mitral or tricuspid valves, or regurgitation of the aortic or pulmonary valves.
* **Continuous murmur.**A heart murmur that occurs throughout the cardiac cycle.

Murmurs related to a congenital (present at birth) heart defect or other problem involving the heart structures will be heard the loudest in the area of the chest where the problem occurs. Some large defects have almost no murmur in the newborn due to normally elevated pressures in the blood vessels in the lungs. Murmurs may be inconsistent and difficult to hear in an infant who is agitated or crying. Thus, murmurs may be missed or not detected.

**Causes**

A heart murmur is caused by rapid, choppy (turbulent) blood flow through the heart. A heart murmur may happen:

* When the heart is filling with blood (diastolic murmur)
* When the heart is emptying (systolic murmur)
* Throughout the heartbeat (continuous murmur)

**Harmless (innocent) heart murmurs**

A person with an innocent murmur usually has a typical heart. Innocent heart murmurs are common in newborns and children.

Things that might change blood flow and cause an innocent heart murmur include:

* Fever
* Lack of healthy red blood cells that carry oxygen to body tissues (anemia)
* Overactive thyroid (hyperthyroidism)
* Phases of rapid growth, such as adolescence
* Physical activity or exercise
* Pregnancy

Innocent heart murmurs may go away over time. Sometimes, heart murmurs continue for life without causing serious health problems.

**Worrisome heart murmurs**

In children, worrisome murmurs are usually due to a problem with the heart's structure that's present at birth (congenital heart defect).

Congenital causes of worrisome heart murmurs include:

* **Holes in the heart.** A hole may form before birth in the wall between the upper or lower heart chambers, or both. Examples are atrial septal defect and ventricular septal defect.
* **Cardiac shunts.** A heart structure problem present before birth (congenital heart defect) causes irregular blood flow between the heart chambers or blood vessels.

In adults, worrisome heart murmurs are usually due to heart valve problems that develop later in life (acquired heart valve disease). Things that can damage the heart valves include:

* **Calcium deposits.** Calcium deposits can cause a heart valve — such as the mitral valve or the aortic valve — to become stiff and narrow (valve stenosis). A calcified valve also may not close completely, letting blood flow backward. The changes in blood flow create a murmur. When calcium affects the heart valves, it's called calcific or degenerative valve disease.
* **Infection of the inner heart lining and valves (endocarditis).** Bacteria or other germs cause this infection. The germs spread through the blood to the heart from another part of the body, such as the mouth. A murmur may occur if the infection is untreated and affects the heart valves.
* **Rheumatic fever.** This is a serious complication of strep throat. It's rare in the United States. Rheumatic fever may occur in those who don't get or complete treatment for strep throat. When rheumatic fever affects the heart valves, it's called rheumatic heart disease. A heart valve problem can cause a murmur.

**Risk factors**

Things that increase the risk of heart murmurs in babies includes:

* Family history of heart problems linked to murmurs
* Uncontrolled diabetes in the mother during pregnancy
* German measles (rubella) in the mother during pregnancy
* Use of certain medications, alcohol or illegal drugs by the mother during pregnancy

Some medical conditions can increase the risk of heart murmurs, including:

* A rare cancerous tumor that releases certain chemicals into the bloodstream (carcinoid syndrome)
* A weakened heart muscle (cardiomyopathy)
* An infection of the lining of the heart (endocarditis)
* Anemia
* Blood disorders marked by a high number of certain white cells, called eosinophils (hypereosinophilic syndrome)
* Certain autoimmune disorders such as lupus and rheumatoid arthritis
* Heart valve disease
* High blood pressure in the lungs (pulmonary hypertension)
* History of rheumatic fever
* Overactive thyroid (hyperthyroidism)

**Symptoms**

Harmless (innocent) heart murmurs usually don't cause any other symptoms.

Symptoms of worrisome heart murmurs depend on the cause. Heart murmur symptoms may include:

* Blue or gray fingernails or lips
* Chest pain
* Cough that doesn't go away
* Dizziness
* Swollen liver
* Swollen neck veins
* Fainting
* Heavy sweating with little or no activity
* In infants, poor appetite and lack of growth
* Shortness of breath
* Swelling or sudden weight gain

**Diagnosis methods**

**Heart Murmur Diagnosis**

Usually, doctors find heart murmurs during a physical exam. Your doctor should be able to hear the sound when listening to your heart with a stethoscope. The doctor will listen for these things:

* How loud is the murmur on a scale of one to six?
* Is the sound high- or low-pitched?
* Where in your heart is the sound? Does it spread to your neck or back?
* Does it happen when your heart pumps, relaxes, or both?
* Does the sound change when you hold your breath, squat, lie down, or stand up?

Other tests your doctor might do during the physical exam are:

* Blood pressure. This test measures the force of your blood pushing against your artery walls. High blood pressure can make the condition that caused your heart murmur worse.
* Blood oxygen level. Anemia can cause your blood oxygen level to drop.
* Pulse rate. A normal resting heart rate is 60-100 beats per minute.

Your doctor may order one or more of these tests to see whether your heart murmur is innocent, or if a valve disease or heart defect caused it:

* Echocardiogram. This test uses sound waves to show the blood flow through your heart. A stress echocardiogram shows how well your heart works when it's under stress from exercise.
* Electrocardiogram (EKG). It measures the electrical activity of your heart.
* Chest X-rays. These images show whether your heart is enlarged due to heart or valve disease.
* Cardiac catheterization. This test shows how well your heart and its blood vessels are working. Your doctor might do cardiac catheterization if they can't find the cause of your murmur. A thin tube goes through a blood vessel to your heart. Sometimes, dye is put into the tube to help your blood vessels show up more clearly on the test.

**Treatment options**

The volume of a heart murmur doesn't always relate to how severe it is. A loud murmur may not be more severe than a quiet one. The type of murmur is a sign of its severity, though. Systolic heart murmurs are often innocent, or harmless.

Your exam and tests should show what caused your heart murmur. If the murmur is related to a heart problem, your doctor may refer you to a heart doctor called a cardiologist. You might need medicine or surgery to treat the cause.

**Treatment for Heart Murmur**

Many children and adults have harmless heart murmurs that don't need treatment. If a condition such as high blood pressure is the cause of your murmur, your doctor will treat the cause.

Treatment for some types of heart valve disease includes medicines such as:

* Blood thinners. These medicines prevent blood clots from forming in the heart.
* Antiarrhythmic medicines. They control a fast or irregular heartbeat.
* Blood pressure lowering medicines. Angiotensin converting enzyme (ACE) inhibitors and beta-blockers lower high blood pressure to prevent it from making your heart murmur worse.
* Water pills. Diuretics remove excess water from your blood so it's easier for your heart to pump.

Some of the heart conditions that cause murmurs need surgery to fix. Your surgeon can do the procedure through a large incision with open-heart surgery. Or you might have minimally invasive or robotic surgery that uses smaller incisions. Sometimes, surgeons perform procedures through a catheter.

Surgery can:

* Close a hole in the heart wall
* Fix or replace a damaged heart valve
* Patch a leak in the valve
* Tighten the ring around a valve so it doesn't leak
* Remove extra tissue to help the valve close tighter

**Prevention**

You can't always prevent heart murmurs. But following a heart-healthy lifestyle and getting regular checkups could help you avoid conditions such as high blood pressure and heart valve problems that cause murmurs.

Not drinking or doing illegal drugs and getting treated for infections during pregnancy can prevent some heart murmurs in children. Most children who do have heart murmurs outgrow them as they get older.

**When to Worry About a Heart Murmur**

Most heart murmurs aren't serious. But sometimes a murmur can be a sign of a valve or heart problem that does need treatment.

Get medical help if you have symptoms such as:

* Chest pain
* Shortness of breath during exercise or when you sleep
* Tiredness
* Dizziness or fainting for no obvious reason
* Heart palpitations
* Swelling in your ankles or legs.

**Prognosis**

In people with innocent heart murmurs, the prognosis is excellent. For people with other types of heart murmurs, the prognosis depends on the type of heart problem and its severity. In general, even when heart surgery is required, the prognosis is good.

**Possible complications**

You are more likely to develop a heart murmur if you have a family history of heart defects or certain medical conditions, including high blood pressure, endocarditis, rheumatoid arthritis, a previous or current case of rheumatic fever, or high blood pressure in the lungs (pulmonary hypertension)

**When to see a doctor / red flag**

Seek medical attention if you have any signs of a heart problem, including:

* Bluish skin.
* Chest pain or pressure.
* Heart palpitations.
* Shortness of breath.
* Syncope or weakness.

**Differential Diagnosis: Heart Sounds & Murmurs**

* Heart Failure
* Ischemic heart disease (anginal equivalent)
* Pulmonary Embolism
* Lung Disease including COPD & Asthma
* Severe Anemia

| **New York Heart Association Classification for Heart Failure** | |
| --- | --- |
| **Class I** | Dyspnea only with vigorous exertion |
| **Class II** | Dyspnea with moderate exertion |
| **Class III** | Dyspnea with mild exertion, may have mild dyspnea at rest |
| **Class IV** | Significant dyspnea at rest |

**Chest pain at rest**

* Myocardial infarction
* Unstable angina
* Dissecting aortic aneurysm
* Esophagitis
* Pulmonary Embolism
* Pneumothorax
* Pericarditis
* Pleuritic pain
* Musculoskeletal pain including costochondritis
* Herpes zoster

**Chest pain on exertion**

* Angina related to atherosclerosis
* Coronary vasospasm with normal coronary arteries
* Aortic stenosis
* Hypertrophic cardiomyopathy
* Musculoskeletal

**Differential Diagnosis of Systolic Murmurs**

**Systolic Ejection**

* **Benign**
  + Innocent systolic murmur (vibratory)
  + Flow murmurs
    - Hemodynamic effect (i.e., fever, hyperthyroidism, severe anemia)
    - Athlete's heart
  + Atrial septal defect (incidental)
* **Pathologic**
  + Aortic stenosis
  + Pulmonary stenosis
  + Hypertrophic cardiomyopathy
  + Atrial septal defect
* **Pansystolic**
  + Tricuspid reflux
  + Mitral reflux
  + Ventricular septal

**Differential Diagnosis of Pericardial Friction Rub**

Caused by inflammation of the pericardial sac with or without fluid.

**Pericarditis**

* **Infectious**
  + viral,
  + bacterial
  + tuberculous
  + fungal
* **Noninfectious**
  + myocardial infarction
  + uremia
  + malignancy
  + myxedema
  + trauma
  + open-heart surgery
* **Autoimmune**
  + rheumatic fever
  + drug induced (procainamide)
  + post-myocardial infarction (Dressler's syndrome)
  + collagen vascular disease

| * + **SYSTOLIC MURMURS** |  |
| --- | --- |
| * + Ejection systolic | * + Flow (thyrotoxicosis, pregnancy, anaemia,) Aortic stenosis Pulmonary stenosis Aortic coarctation Hypertrophic cardiomyopathy |
| * + Pan systolic | * + Mitral regurgitation Ventricular septal defect Tricuspid regurgitation |
| * + Late systolic | * + Mitral valve prolapse |
| * + Early systolic | * + Acute severe mitral regurgitation |
| * + **DIASTOLIC MURMURS** |  |
| * + Early diastolic | * + Aortic regurgitation Pulmonary regurgitation |
| * + Mid-late diastolic | * + Mitral stenosis Austin Flint |

**Epidemiology data**

Of the 1764 pupils recruited, 900 (51.02%) were females while 864 (48.98%) were males. The mean age of the pupils was 8.86 ± 2.14 years. 1065 (60.37%) and 699 (39.63%) respectively were recruited from public and private schools. Of the 1764 pupils, six (0.34%) had significant murmur. Only one of the six had RHD, giving a prevalence of 0.57/1000 pupils. The pupil with RHD was a male, from public school and in the low socioeconomic class.

The prevalence of RHD in this study is low compared to similar studies conducted outside the country. The true prevalence may be underestimated since higher prevalence is obtained from echocardiographic based screening compared to clinical screening.

In neonates, heart murmurs are detected in a small percentage of cases. A study involving 6,333 healthy newborns found that murmurs were detected in 87 babies, representing a prevalence of 1.37%.5 Among these, 37 babies (42.5%) had structural cardiac malformations, with ventricular septal defect being the most common diagnosis.5 This indicates that while many murmurs are innocent and harmless, there is a significant chance that a detected murmur could indicate an underlying structural defect, necessitating further investigation.5

In children, approximately 50-70% of heart murmurs are clinically insignificant.5 The prevalence of heart murmurs in neonates varies widely, from 0.6% to 77.4%, with most reports coming from early studies before the advent of echocardiography and interventional cardiology, which have improved the accuracy of diagnosing congenital heart disease.5

For routine neonatal examinations, if a murmur is detected, it should prompt early referral for investigation and diagnosis or appropriate family reassurance.5 This is crucial as heart murmurs are the most common reason for referral to a pediatric cardiologist.5

In summary, while the specific epidemiological data for heart murmurs in Nigeria is limited, studies suggest that murmurs are relatively uncommon in neonates and children, with a significant proportion being innocent. However, the possibility of underlying structural heart defects necessitates careful clinical evaluation and, if necessary, further diagnostic testing

**References**

[**https://www.health.harvard.edu/a\_to\_z/heart-murmur-a-to-z**](https://www.health.harvard.edu/a_to_z/heart-murmur-a-to-z)

[**https://my.clevelandclinic.org/health/diseases/17083-heart-murmur**](https://my.clevelandclinic.org/health/diseases/17083-heart-murmur)

[**https://depts.washington.edu/physdx/heart/diffdx.html**](https://depts.washington.edu/physdx/heart/diffdx.html)

[pubmed.ncbi.nlm.nih.gov/33342586/](https://pubmed.ncbi.nlm.nih.gov/33342586/).

Swartz MH. The heart. In: Swartz MH, ed. *Textbook of Physical Diagnosis: History and Examination.* 8th ed. Philadelphia, PA: Elsevier; 2021:chap 14.

<https://www.webmd.com/heart-disease/heart-murmur-causes-treatments>

<https://www.mayoclinic.org/diseases-conditions/heart-murmurs/symptoms-causes/syc-20373171>

**Myocardial Infarction (Heart Attack)**

Sudden blockage of blood flow to the heart

**What is a heart attack?**

A heart attack (myocardial infarction) is an extremely dangerous condition that happens because you don’t have enough blood flow to some of your heart muscle. This lack of blood flow can occur because of many different factors but is usually related to a blockage in one or more of your heart’s arteries.

Without blood flow, the affected heart muscle will begin to die. If you don’t get blood flow back quickly, a heart attack can cause permanent heart damage and/or death.

A heart attack is a life-threatening emergency. Time is critical in treating a heart attack. A delay of even a few minutes can result in permanent heart damage or death.

**What exactly happens during a heart attack?**

When a heart attack happens, blood flow to a part of your heart stops or is far below normal, which causes injury or death to that part of your heart muscle. When a part of your heart can’t pump because it’s dying from lack of blood flow, it can disrupt the pumping function of your heart. This can reduce or stop blood flow to the rest of your body, which can be deadly if someone doesn’t correct it quickly.

A heart attack occurs when one of the heart's coronary arteries is blocked suddenly or has extremely slow blood flow. A heart attack also is called a myocardial infarction.

The usual cause of sudden blockage in a coronary artery is the formation of a blood clot (thrombus). The blood clot typically forms inside a coronary artery that already has been narrowed by atherosclerosis, a condition in which fatty deposits (plaques) build up along the inside walls of blood vessels.

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Slow blood flow in a coronary artery can happen when the heart is beating very fast or the person has low blood pressure. If the demand for oxygen is greater than the supply, a heart attack can happen without formation of a blood clot. People with atherosclerosis are also more likely to have this reason for a heart attack.

Each coronary artery supplies blood to a specific part of the heart's muscular wall, so a blocked artery causes pain and malfunction in the area it supplies. Depending on the location and amount of heart muscle involved, this malfunction can seriously interfere with the heart's ability to pump blood. Also, some of the coronary arteries supply areas of the heart that regulate heartbeat, so a blockage sometimes causes potentially fatal abnormal heartbeats, called cardiac arrhythmias.

The pattern of symptoms that develops with each heart attack and the chances of survival are linked to the location and extent of the coronary artery blockage.

Most heart attacks result from atherosclerosis. The risk factors for heart attack and atherosclerosis are basically the same:

* an abnormally high blood level of LDL (low-density lipoprotein) cholesterol, triglycerides, and/or total cholesterol (hypercholesterolemia)

**Causes and risk factors**

The blockage is caused by a buildup of plaque in the arteries (atherosclerosis). Plaque is made up of deposits, cholesterol, and other substances. When a plaque breaks (ruptures), a blood clot quickly forms. The blood clot is the actual cause of the heart attack.

If the blood and oxygen supply is cut off, muscle cells of the heart begin to suffer damage and start to die. Irreversible damage begins within 30 minutes of blockage. The result is heart muscle affected by the lack of oxygen no longer works as it should.

These groups are most at risk:

* People with inherited high blood pressure (hypertension)
* People with inherited low levels of HDL cholesterol, high levels of LDL cholesterol, or high levels of triglycerides
* People with a family history of heart disease. This is especially true if the heart disease started before age 55.
* Older men and women
* People with type 1 diabetes
* Women who have gone through menopause. Generally, men are at risk at a younger age than women. After menopause, women are equally at risk.

**Acquired risk factors: Who is most at risk?**

These groups are most at risk:

* People with acquired high blood pressure (hypertension)
* People with acquired low levels of HDL cholesterol, high levels of LDL cholesterol, or high levels of triglycerides
* Cigarette smokers
* People who are under a lot of stress
* People who drink too much alcohol
* People who lead a sedentary lifestyle
* People overweight by 30% or more
* People who eat a diet high in saturated fat
* People with type 2 diabetes

A heart attack can happen to anyone. When you take the time to learn which risk factors apply to you, you can take steps to eliminate or reduce them.

**Signs and Symptoms**

Many people feel pain in their chest during a heart attack. It can feel like discomfort, squeezing or heaviness, or it can feel like crushing pain. It may start in your chest and spread (or radiate) to other areas like your left arm (or both arms), shoulder, neck, jaw, back or down toward your waist.

People often think they’re having indigestion or heartburn when they’re actually having a heart attack.

Some only experience shortness of breath, nausea or sweating.

**What are the symptoms of a heart attack?**

Heart attacks can have many symptoms, some of which are more common than others.

Heart attack symptoms that people describe most often include:

* Chest pain (angina)
  + Sweating, cool, clammy skin, or paleness
* Severe pressure, fullness, squeezing, pain, or discomfort in the center of the chest that lasts for more than a few minutes
* Pain or discomfort that spreads to the shoulders, neck, arms, or jaw
* Chest pain that gets worse
* Chest pain that doesn't get better with rest or by taking nitroglycerin
* Shortness of breath or trouble breathing
* Trouble sleeping (insomnia)
* Nausea or stomach discomfort
* Heart palpitations
* Anxiety or a feeling of “impending doom”
* Feeling lightheaded, dizzy or passing out

Unexplained weakness or fatigue

Rapid or irregular pulse

Dizziness

Men are likely to have different heart attack symptoms. Women are less likely to have chest pain or discomfort that feels like indigestion. They’re more likely to have shortness of breath, fatigue and insomnia that started before the heart attack.

Although chest pain is the key warning sign of a heart attack, it may be confused with other conditions. These include indigestion, pleurisy, pneumonia, tenderness of the cartilage that attaches the front of the ribs to the breastbone, and heartburn. Always see your healthcare provider for a diagnosis.

**Diagnosis methods**

Ideally, a health care provider should screen you during regular checkups for risk factors that can lead to a heart attack.

A heart attack is often diagnosed in an emergency setting. If you've had or are having a heart attack, care providers will take immediate steps to treat your condition. If you're able to answer questions, you may be asked about your symptoms and medical history.

Diagnosis of a heart attack includes checking blood pressure, pulse and temperature. Tests are done to see how the heart is beating and to check overall heart health.

**Tests**

Tests to diagnose a heart attack include:

* **Electrocardiogram (ECG or EKG).** This first test done to diagnose a heart attack records electrical signals as they travel through the heart. Sticky patches (electrodes) are attached to the chest and sometimes the arms and legs. Signals are recorded as waves displayed on a monitor or printed on paper. An electrocardiogram (ECG) can show if you are having or have had a heart attack.
* **Blood tests.** Certain heart proteins slowly leak into the blood after heart damage from a heart attack. Blood tests can be done to check for these proteins (cardiac markers).
* **Chest X-ray.** A chest X-ray shows the condition and size of the heart and lungs.
* **Echocardiogram.** Sound waves (ultrasound) create images of the moving heart. This test can show how blood moves through the heart and heart valves. An echocardiogram can help identify whether an area of your heart has been damaged.
* **Coronary catheterization (angiogram).** A long, thin tube (catheter) is inserted into an artery, usually in the leg, and guided to the heart. Dye flows through the catheter to help the arteries show up more clearly on images made during the test.
* **Cardiac computed tomography (CT) or Magnetic resonance imaging (MRI).** These tests create images of the heart and chest. Cardiac CT scans use X-rays. Cardiac MRI uses a magnetic field and radio waves to create images of your heart. For both tests, you usually lie on a table that slides inside a long tubelike machine. Each test can be used to diagnose heart problems. They can help show the severity of heart damage.

**Treatment options**

Treatment for a heart attack

The goal of treatment for a heart attack is to relieve pain, preserve the heart muscle function, and prevent death.

Treatment in the emergency department may include:

* Intravenous therapy, such as nitroglycerin and morphine
* Continuous monitoring of the heart and vital signs
* Oxygen therapy to improve oxygenation to the damaged heart muscle
* Pain medicine to decrease pain. This, in turn, decreases the workload of the heart. The oxygen demand of the heart decreases.
* Cardiac medicine such as beta-blockers to promote blood flow to the heart, improve the blood supply, prevent arrhythmias, and decrease heart rate and blood pressure
* Fibrinolytic therapy. This is the intravenous infusion of a medicine that dissolves the blood clot, restoring blood flow.
* Antithrombin or antiplatelet therapy with aspirin or clopidogrel. This is used to prevent further blood clotting.
* Antihyperlipidemics. These medicines lower lipids (fats) in the blood, particularly low density lipid (LDL) cholesterol. Statins are a group of antihyperlipidemic medicines. They include simvastatin, atorvastatin, and pravastatin. Bile acid sequestrants—colesevelam, cholestyramine, and colestipol—and nicotinic acid (niacin) are two other types of medicines that may be used to lower cholesterol levels.

You may need other procedures to restore blood flow to the heart. Those procedures are described below.

**Coronary angioplasty**

With this procedure, a balloon is used to create a bigger opening in the vessel to increase blood flow. This is often followed by inserting a stent into the coronary artery to help keep the vessel open. Although angioplasty is done in other blood vessels elsewhere in the body, percutaneous coronary intervention (PCI) refers to angioplasty in the coronary arteries. This lets more blood flow into the heart. PCI is also called percutaneous transluminal coronary angioplasty (PTCA). There are several types of PTCA procedures:

* **Balloon angioplasty.** A small balloon is inflated inside the blocked artery to open the blocked area.
* **Coronary artery stent.** A tiny coil is expanded inside the blocked artery to open the blocked area. The stent is left in place to keep the artery open.
* **Atherectomy.**The blocked area inside the artery is cut away by a tiny device on the end of a catheter.
* **Laser angioplasty.** A laser used to "vaporize" the blockage in the artery.

**Coronary artery bypass**

This surgery is most commonly referred to as simply bypass surgery or CABG (pronounced "cabbage"). It is often done in people who have chest pain (angina) and coronary artery disease. Coronary artery disease is when plaque has built up in the arteries. During the surgery, the surgeon makes a bypass by grafting a piece of a vein above and below the blocked area of a coronary artery. This lets blood flow around the blockage. The surgeon usually takes veins from a leg, but he or she may also use arteries from the chest or an arm. Sometimes, you may need more than one bypass surgery to restore blood flow to all areas of the heart.

**Management and Treatment**

**How is a heart attack treated?**

* Treating a heart attack means restoring blood flow to the affected heart muscle as soon as possible. This can happen in a variety of ways, ranging from medication to surgery. Treatment will likely include several of the following methods.
* **Supplementary oxygen**
* People having trouble breathing or with low blood oxygen levels often receive supplementary oxygen along with other heart attack treatments. You can breathe the oxygen either through a tube that sits just below your nose or a mask that fits over your nose and mouth. This increases the amount of oxygen circulating in your blood and reduces the strain on your heart.

**Medications**

These may include:

* **Anti-clotting medications.** This includes aspirin and other blood-thinning medicines.
* **Nitroglycerin.** This medicine relieves chest pain and causes blood vessels to widen so blood can pass through more easily.
* **Thrombolytic (clot-busting) medications.** Providers use these only within the first 12 hours after a heart attack.
* **Antiarrhythmia medications.** Heart attacks can often cause malfunctions in your heart’s normal beating rhythm called arrhythmias, which can be life-threatening. Antiarrhythmic medications can stop or prevent these malfunctions.
* **Pain medications.** The most common pain medication people receive during heart attack care is morphine. This can help alleviate chest pain.
* **Beta-blockers.** These medications help slow down your heart rate so your heart can recover from the injury of a heart attack.
* **Antihypertensives.** These medications decrease your blood pressure and can help your heart recover from the injury of a heart attack.
* **Statins.** These medications help stabilize the plaque in your heart’s blood vessels so they’re less likely to rupture. They also help reduce cholesterol and the chances of having another heart attack.
* **Percutaneous coronary intervention**
* Providers restore circulation to your affected heart muscle with a procedure called percutaneous coronary intervention (PCI) or angioplasty.
* Opening your artery with a catheter is critical in restoring blood flow. The sooner that happens, the better the chance of a good outcome. Providers use a metric called “door-to-balloon time” to measure their ability to treat a heart attack. This is the average time it takes for people to undergo PCI after they first come into the emergency room. If you receive PCI, your provider may place a stent at the site of the blockage. The stent helps hold your artery open so another blockage doesn’t happen in the same spot.
* **Coronary artery bypass grafting**
* People who have severe blockages of their coronary arteries may undergo coronary artery bypass grafting (CABG). Providers often call this open-heart surgery or bypass surgery.
* CABG involves using a blood vessel from elsewhere in your body (usually your chest, arm or leg) to construct a detour for blood. This reroutes blood around one or more blocked artery sections and brings blood to your heart muscle.

**Complications/side effects of the treatment**

* Treatment side effects vary by treatment and may include:
* Nausea and vomiting
* Weakness
* Lightheadedness
* Bleeding
* Infection
* Damage to a blood vessel
* Arrhythmia
* Kidney issues
* Stroke

**How soon after treatment will I feel better?**

* In general, your heart attack symptoms should decrease as you receive treatment. You’ll likely have some lingering weakness and fatigue during your hospital stay and for several days after. Your healthcare provider will give you guidance on rest, medications to take and more.
* Recovery from the treatments also varies, depending on the method of treatment. The average hospital stay for a heart attack is between four and five days. In general, expect to stay in the hospital for the following length of time:
* **Medication only.** People who only receive medication have an average hospital stay of about six days.
* **PCI.** Recovering from PCI is easier than surgery because it’s a less invasive method for treating a heart attack. The average length of stay for PCI is about four days.
* **CABG.** Recovery from heart bypass surgery takes longer because it’s a major surgery. The average length of stay for CABG is about eight to 12 days.

**Prevention**

In general, there are many things you can do that may prevent a heart attack. However, there are some factors you can’t change — especially your family history — that can still lead to a heart attack despite best efforts. Still, reducing your risk can postpone when you have a heart attack and reduce the severity if you do have one.

**How can I lower my risk?**

Although several risk factors can’t be modified, there are many ways you can help yourself and reduce your risk of a heart attack, including:

* **Scheduling a check-up.** Find a primary care provider (PCP) and see them at least once a year for a check-up or wellness visit. An annual check-up can catch many of the early warning signs of heart disease, including changes that you can’t feel. These include your blood pressure, blood sugar levels, cholesterol levels and more.
* **Quitting tobacco products.** This includes smokeless tobacco and all vaping products.
* **Exercising regularly.** Aim for 30 minutes of moderately intense physical activity five days a week.
* **Eating nutritious foods.** Examples include the Mediterranean or Dash diets. Eating plant-based meals is an excellent alternative to eating lots of processed meats and saturated fats.
* **Maintaining a weight that’s healthy for you.** Your primary care provider can advise you on a healthy target weight and provide resources and guidance to help you reach that goal.
* **Managing your existing health conditions.** This includes high cholesterol levels, high blood pressure and diabetes.
* **Reducing your stress.** Consider techniques such as yoga, deep breathing and meditation.
* **Taking your medications as prescribed.** Don’t just take medications when you remember to or when you have a doctor’s appointment coming up.
* **Keeping all of your medical appointments.** Seeing your healthcare providers regularly can help uncover heart-related issues or other medical problems you didn’t know you had. This can also help treat problems sooner rather than later.

Being an active contributor to your health doesn’t mean you have to make lifestyle changes all on your own. Ask your primary care provider and others on your healthcare team for help. They can provide the information and resources you need.

If you’ve already had a heart attack, your healthcare provider will recommend a cardiac rehabilitation program. This program’s goal is to reduce your chance of a second heart attack. These medically supervised programs provide counseling and focus on the same healthy living goals listed above.

**Outlook / Prognosis**

Survival from a heart attack has improved dramatically over the last two decades. However, some people experience sudden death and never make it to the hospital. For most people that do reach the hospital soon after the onset of symptoms, the prognosis is very good. Many people leave the hospital feeling well with limited heart damage.

The treatment you receive for a heart attack doesn’t end when you leave the hospital. Your risk of a second heart attack means you’ll need to take prescribed medicines and make changes to your daily life.

**When can I resume my usual activities?**

Recovery from a heart attack after leaving the hospital depends on the severity of the heart attack, how soon treatment began, which treatments you had and the health conditions you had — if any — before your heart attack.

Your healthcare provider can explain the next steps for your recovery and what you can expect. In general, most people can return to work or resume their usual activities anywhere between two weeks to three months after their heart attack. Cardiac rehab can help people gradually and safely increase their physical activity back to its prior level.

**Outlook for a heart attack**

Today, many people survive a heart attack. But the outlook is worse for those who:

* Are over 65
* Have heart failure
* Have diabetes
* Already had a heart attack

They may be at risk of another heart attack or of dying in the next six months or less. In these cases, providers will work to aggressively manage their risks.

Premenopausal women under age 45 have a better outcome than men of a similar age. Scientists believe this is because of estrogen’s heart-protective effects. But after menopause ends the protective benefits of estrogen, females fare worse than males. More specifically:

* Women between the ages of 45 and 65 who’ve had a heart attack are more likely to die within a year of the event compared with men of this same age.
* Women over age 65 are more likely to die within weeks of their heart attacks than men over age 65.

**How do I take care of myself?**

After a heart attack, you’ll continue to take medicines — some of which you received for immediate treatment of your heart attack — long term. These include:

* Beta-blockers
* ACE inhibitors
* Aspirin and other blood-thinning agents
* Statins

**Possible complications**

Complications associated with heart attacks include:

* Arrhythmias (abnormal heart rhythms)
* Heart failure
* Heart valve problems
* Stroke
* Sudden cardiac arrest
* Depression and anxiety
* Cardiogenic shock
* Mechanical complications of a heart attack, such as a ventricular septal defect or free wall rupture (these are more likely to happen with delayed treatment of a heart attack)

**When to see a doctor / red flag**

After you’ve had a heart attack, you’re at a higher risk of a similar occurrence. Your healthcare provider will likely recommend follow-up monitoring, testing and care to avoid future heart attacks. Some of these include:

* **Heart scans.** Like the methods providers use to diagnose a heart attack, these can assess the effects of your heart attack and determine if you have permanent heart damage. They can also look for signs of heart and circulatory problems that increase the chance of future heart attacks.
* **Stress test.** These heart tests and scans that take place while you’re exercising can show potential problems that stand out only when your heart is working harder.
* **Cardiac rehabilitation.** These programs help you improve your overall health and lifestyle, which can prevent another heart attack.

**When should I go to the ER?**

If you have some of the heart attack symptoms listed here and think you’re having a heart attack, call your local emergency number. While they send help, ask if you should take an aspirin or other medicine. Don’t wait to call. Minutes matter when you’re having a heart attack

**What questions should I ask my doctor?**

Questions to ask your healthcare provider may include:

* What activities are safe for me to do after a heart attack?
* What are the most important things I should do to prevent another heart attack?
* How often do I need follow-up appointments with you?
* Should others in my family get heart check-ups?

**Differential diagnosis**

Due to the vague nature of chest pain, it can be representative of a wide range of medical conditions. differential diagnosis can be broken down into categories:

* **Cardiac**: acute coronary syndrome, pericarditis, congestive heart failure, post-cardiac injury syndrome
* **Hematologic**: sickle cell anemia, pleural effusion
* **Gastrointestinal**: pancreatitis, inflammatory bowel disease, bacterial pleurisy
* **Infections**: abscesses to the liver, lung, or spleen, bacterial and viral infections

Other categories of differential diagnosis are iatrogenic, pulmonary, rheumatologic, and renal.

**Myocardial Infarction Differential Diagnoses**

Myocardial infarction, commonly known as a heart attack, can be differentiated from other conditions through a variety of diagnostic criteria and tests. Here are some conditions that can present similarly to myocardial infarction but are distinct:

* **Acute Pericarditis**: This condition involves inflammation of the pericardium, the membrane surrounding the heart. Symptoms include sharp chest pain, tachycardia, and elevated troponin levels. However, acute pericarditis typically presents with widespread ST-segment elevation on EKG, unlike the localized ST-segment elevation seen in myocardial infarction.
* **Stable Angina**: Stable angina is characterized by chest pain that occurs during exertion due to insufficient blood flow to the heart muscle. Unlike myocardial infarction, stable angina resolves with rest and does not result in myocardial cell necrosis. Patients with stable angina do not have elevated troponin levels or ST elevation on ECG.
* **Pulmonary Embolism**: This condition involves a blockage in the pulmonary arteries, often due to blood clots. Symptoms can include chest pain, shortness of breath, and elevated troponin levels. However, pulmonary embolism does not typically cause ST-segment elevation on EKG, which is a hallmark of myocardial infarction.
* **Gastroesophageal Reflux Disease (GERD)**: GERD can cause chest pain that mimics myocardial infarction. However, the pain from GERD is usually relieved by antacids and does not present with ECG changes or elevated troponin levels characteristic of a heart attack.
* **Musculoskeletal Pain**: Pain originating from the muscles or bones in the chest area can mimic cardiac pain. This type of pain is typically sharp and localized and does not cause ECG changes or elevated troponin levels.

**Epidemiology data**

The health infrastructures 5th contributors to age-specific mortality. Major adverse cardiac events following acute coronary events are seen in 30.8%, and in-hospital mortality is 8.1%. Mortality rates at 30 days, 3 months, 6 months, and 1 year were 8.7%, 9.9%, 10.9%, and 13.3%, respectively. For heart failure, the mortality rate at 1 year is 12.3%. From a current-to-future health perspective, hypertension, cardiomyopathies, and rheumatic disease are the most common forms of CVD. Moreover, Nigeria has the highest global burden of peripartum cardiomyopathy. The 30.6% prevalence of hypertension is of major concern. Only 29.0% of people are aware of their hypertension, 12.0% are receiving treatment, and 2.8% had at-goal blood pressure in 2020.

Nigeria has historically lacked local guidelines for managing CVD. Clinicians therefore depend on the guidelines in North America and Europe. However, national guidelines exist for managing hypertension; pharyngitis, acute and chronic rheumatic heart disease, and heart disease within the COVID pandemic context.

In 2019, Nigeria launched a national multisectoral action plan for the prevention and control of noncommunicable diseases (including CVD). A unit at the Federal Ministry of Health now oversees the control of noncommunicable diseases with national surveillance and monitoring systems. The country’s Tobacco Control Act was enacted in 2015. A National Health Bill was signed into law that includes provision of funds and essential medicines for common diseases including CVD. The effect of these initiatives has not been assessed.

Nigerians are now at risk of developing coronary artery disease. The burden of hypertension is high, and poverty is a major and constant problem and threat to controlling unhealthy behaviors and diets and providing access to quality health care. Nigeria unfortunately has not fully implemented the national Tobacco Control Act, and although there is an Essential Medicines List, these drugs are often unavailable in public health facilities. With the devaluation of the country’s currency, importation of medicines and devices has become extremely expensive.

Experts say heart diseases, also known as cardiovascular diseases (CVDs), are a major contributor to the cases of sudden deaths or people slumping and dying in the country.

According to Dr Charles Anjorin, a consultant physician and cardiologist at the University of Maiduguri Teaching Hospital, in Borno State, cardiovascular diseases are most implicated in cases of sudden deaths, which have become very frequent in Nigeria.

“By sudden death, we mean unexpected death in an individual previously thought to be quite well. Common causes of this include severe hypertension, acute heart failure, stroke, heart attack (myocardial infarction) and disorders of the electrical activity of the heart, also known as disorders of heart rhythm, “ he said.

Nigerians and Africans are shifting away from the agrarian rural lifestyles and diets and when people walked long distances to go to the farm or school.

REFERENCES

[**Heart diseases spike, fuel sudden deaths in Nigeria - Daily Trust**](https://dailytrust.com/heart-diseases-spike-fuel-sudden-deaths-in-nigeria/)

**National Heart, Lung, and Blood Institute (NHLBI)**  
[https://www.nhlbi.nih.gov/](http://www.nhlbi.nih.gov/)

**American Heart Association (AHA)**  
[https://www.heart.org/](http://www.heart.org/)

<https://www.hopkinsmedicine.org/health/conditions-and-diseases/heart-attack>

<https://my.clevelandclinic.org/health/diseases/16818-heart-attack-myocardial-infarction>

**Rheumatic Heart Disease**

**Definition and description**

Rheumatic heart disease is heart valve damage from rheumatic fever. Bacterial infections called group A streptococcal (GAS) infections can cause rheumatic fever. An infection like strep throat or scarlet fever triggers your body’s immune response, causing inflammation throughout your body, including in your heart.

Inflammation from rheumatic fever can lead to permanent damage to your heart valves. Your valves keep your blood flowing in the right direction through your heart. Damaged valves reduce the amount of blood that can move through your heart. They also may allow some blood to go in the wrong direction.

**How common is rheumatic heart disease?**

Rheumatic heart disease is rare in the U.S. It’s more common in low-income or developing parts of the world where people can’t get antibiotics for bacterial infections. About 300,000 people worldwide die of rheumatic heart disease each year. More than 40 million people in the world have the disease.

Children and teenagers with untreated strep infections are the most likely to get rheumatic fever, often between ages 5 and 15. Signs of heart damage can develop years after the infection and fever are gone. People often show signs of rheumatic heart disease as young adults.

**Causes and risk factors**

**What causes rheumatic heart disease?**

Heart valve inflammation from rheumatic fever causes rheumatic heart disease. The damage may happen right away. Or it can develop over time from repeated strep infections. Continuing inflammation leads to heart valve scarring and narrowing.

**Is rheumatic heart disease contagious?**

Rheumatic heart disease isn’t contagious. But strep throat is. This infection can lead to rheumatic fever, the cause of rheumatic heart disease.

**What are the risk factors for rheumatic heart disease?**

People are at a higher risk for this disease if they:

* Don’t have easy access to healthcare or antibiotics.
* Have repeated strep infections that go untreated.
* Live in overcrowded or unsanitary conditions.

**Signs and symptoms**

Some people may have symptoms of heart involvement during an acute episode of rheumatic fever. But in most cases, rheumatic heart disease symptoms may not appear until years after a strep infection or rheumatic fever. People with heart damage may experience:

* Chest pain.
* Fatigue.
* Heart murmur.
* Shortness of breath with exercise, at rest or when lying flat.
* Swelling in your stomach, hands or feet.
* Palpitations or cardiac arrhythmias like atrial fibrillation.
* Coughing up blood.

Rheumatic fever symptoms can include:

* fever
* painful joints especially knees ankles, elbows and wrists
* pain that moves between different joints
* jerky uncontrollable body movements called chorea
* painless nodules under the skin near joints and/or a rash consisting of pink rings with a clear centre (both rare)

Symptoms of heart valve damage that is associated with rheumatic heart disease may include:

* chest pain or discomfort
* shortness of breath
* swelling of the stomach, hands or feet
* fatigue
* rapid or irregular heartbeat.

**Diagnosis methods**

People with rheumatic heart disease will have or recently had a strep infection. A throat culture or blood test may be used to check for strep.

They may have a murmur or rub that may be heard during a routine physical exam. The murmur is caused by blood leaking around the damaged valve. The rub is caused when the inflamed heart tissues move or rub against each other.

Along with a complete health history and physical exam, tests used to diagnose rheumatic heart disease may include:

* **Echocardiogram (echo).**This test uses sound waves to check the heart's chambers and valves. The echo sound waves create a picture on a screen as a handheld ultrasound probe (transducer) is passed over the skin over the heart. Echo can show damage to the valve flaps, backflow of blood through a leaky valve, fluid around the heart, and heart enlargement. It’s the most useful test for diagnosing heart valve problems. For more in-depth pictures, you may be given sedation, and the probe is put into the throat (transesophageal echo or TEE).
* **Electrocardiogram (ECG).**This test records the strength and timing of the heart's electrical activity. It shows abnormal rhythms (arrhythmias or dysrhythmias). And it can sometimes find heart muscle damage. Small sensors are taped to your skin to pick up the electrical activity.
* **Chest X-ray.**An X-ray may be done to check your lungs and see if your heart is enlarged.
* **Cardiac MRI.** This is an imaging test that takes detailed pictures of the heart. It may be used to get a more exact look at the heart valves and heart muscle.
* **Blood tests.**Certain blood tests may be used to look for infection and inflammation.

**Treatment options**

Rheumatic heart disease treatments can help you manage symptoms and may delay disease progress. But they can’t cure the condition. Treatments include:

* **Medication:**Your provider may recommend medication to manage heart failure or an abnormal heartbeat. Anticoagulants (blood thinners) can reduce the risk of stroke or blood clots if you have a narrow mitral valve or atrial fibrillation.
* **Minimally invasive procedure:** If your mitral valve is narrow but you don’t have other issues with it, a provider can perform a valvuloplasty to widen the valve.
* **Surgery:**If you have severe rheumatic heart disease, you may need heart valve surgery. A surgeon repairs or replaces damaged heart valves. If they can’t fix your valve, they can replace the damaged valve with an artificial valve or a tissue valve. In some cases, they may perform a Ross procedure, where a surgeon swaps one of your healthy valves for the damaged valve and then puts a new valve in place of the healthy one they relocated.

**Prevention tips**

Since rheumatic heart disease results from rheumatic fever, an important strategy is to prevent rheumatic fever from occurring. Treatment of strep throat with appropriate antibiotics will prevent rheumatic fever.

Once a patient has been identified as having had rheumatic fever, it is important to prevent additional streptococcal infections as this could cause a further episode of rheumatic fever and additional damage to the heart valves. The strategy to prevent additional streptococcal infection is to treat the patient with antibiotics over a long period of time. The antibiotic treatment that is most effective in preventing further infection is benzathine penicillin G, which is given by intramuscular injection every 3–4 weeks over many years.

For countries where rheumatic heart disease is endemic, the main strategies for prevention, control and elimination include

* improving standards of living;
* expanding access to screening and appropriate care for people with suspected or confirmed streptococcal infections and RF/RHD and treatment of RHD complications with medications;
* ensuring a consistent supply of quality-assured antibiotics for primary and secondary prevention; and
* planning, developing and implementing feasible programs for prevention and control of rheumatic heart disease, supported by adequate monitoring and surveillance, as an integrated component of national health systems responses.

**Outlook / Prognosis**

The right treatments may delay or prevent heart failure in people with rheumatic heart disease. But the disease is permanent and requires long-term care. Without regular checkups, rheumatic heart disease can lead to severe heart failure.

The length of time that you can live with rheumatic heart disease depends on how severe it is at diagnosis. In a study of indigenous people in Australia under 25, those who had severe rheumatic heart disease at diagnosis got worse fairly quickly. Of this group, 50% had surgery within two years and 10% of them died within six years after their diagnosis.

Some people with moderate disease improved, while others stayed the same or got worse. Ten years after diagnosis, 60% of people with mild rheumatic heart disease didn’t get worse.

**How do I take care of myself?**

If you have rheumatic heart disease, you should avoid getting rheumatic fever again. It can make your heart disease worse. Your provider can monitor your health to make sure you get antibiotics if you get strep throat again. You may also be able to receive antibiotics to prevent the strep infection that causes rheumatic fever.

Follow your provider’s instructions for treating rheumatic heart disease. That may include taking medicines, going to follow-up appointments or having surgery.

**Possible complications**

Rheumatic fever can affect your heart about 20 or 30 years after an episode of rheumatic fever. If you had repeated episodes or were younger when you had rheumatic fever, you may see the consequences of rheumatic heart disease at a younger age.

One or more of your heart valves can grow narrow (referred to as stenosis of the valve) or allow blood to flow backward in the wrong direction. Providers call this regurgitation. Rheumatic heart disease tends to affect the mitral and aortic heart valves.

Rheumatic heart disease can lead to:

* Arrhythmia (abnormal heart rhythms, like atrial fibrillation).
* Heart failure.
* Infective endocarditis, an infection in your heart valves.
* Pulmonary hypertension.

Some of these conditions can increase your risk of stroke or blood clots.

Rheumatic heart disease is especially dangerous if you're pregnant. Pregnancy increases the amount of blood in your body. Your heart has to work harder to pump the extra blood. As a result, a woman with damaged heart valves can have serious health issues during pregnancy. The fetus’s health is also at risk.

**When to see a doctor / red flag**

Contact your provider if you experience new or worsened symptoms, including:

* Confusion.
* Difficulty breathing.
* Chest pain.
* Swelling or pain in your lower body.
* Coughing up blood.

**What questions should I ask my healthcare provider?**

Questions to ask your healthcare provider may include:

* How severe is my case of rheumatic heart disease?
* Did the disease affect one or more of my heart valves?
* Which treatment is best for me?
* Do I need antibiotics to prevent further episodes of rheumatic fever?

**Differential Diagnoses**

* Acute Complications of Sarcoidosis
* Appendicitis Imaging
* Bicuspid Aortic Valve

Carditis

Aortic insufficiency

* Carnitine Deficiency
* Coccidioidomycosis and Valley Fever
* Congenital Mitral Stenosis
* Dilated Cardiomyopathy (DCM)
* Glomerulonephritis
* Heart Failure

Carditis

* Histoplasmosis
* Kawasaki Disease

Carditis

Rash

* Pediatric Aortic Regurgitation (Aortic Valve Insufficiency)

Carditis

Aortic insufficiency

* Pediatric Bacterial Endocarditis

Carditis

Valve insufficiency

* Pediatric Cardiac Tumors
* Pediatric Dilated Cardiomyopathy (DCM)

Carditis

* Pediatric HIV Infection
* Pediatric Infective Pericarditis
* Pediatric Malignant Pericardial Effusion
* Pediatric Mitral Regurgitation (Mitral Valve Insufficiency)

Carditis

Mitral insufficiency

* Pediatric Mitral Valve Prolapse
* Pediatric Non Viral Myocarditis

Carditis

* Pediatric Septic Arthritis
* Pediatric Valvar Aortic Stenosis
* Pediatric Viral Myocarditis

Carditis

* Systemic Lupus Erythematosus (SLE)
* Transient Synovitis

## Acute Complications of Sarcoidosis

* Sarcoidosis is a multisystem granulomatous disease that can acutely involve the lungs, heart, eyes, and nervous system.
* Acute complications include:
  + Pulmonary: Acute respiratory failure due to extensive granulomatous inflammation or fibrosis.
  + Cardiac sarcoidosis: Can cause acute myocarditis, arrhythmias, heart block, or sudden cardiac death.
  + Neurosarcoidosis: Acute cranial nerve palsies or meningeal inflammation.
  + Hypercalcemia: Due to increased vitamin D activation by granulomas, leading to acute kidney injury.

## Appendicitis Imaging

* Imaging (ultrasound, CT scan) is used to confirm diagnosis, identify complications like perforation or abscess.

Bicuspid Aortic Valve

* A congenital valve abnormality with two leaflets instead of three.
* Can cause aortic stenosis or insufficiency, leading to heart failure if untreated.

Carditis (General)

* Inflammation of the heart layers (myocarditis, pericarditis, endocarditis).
* Can lead to arrhythmias, heart failure, or valve dysfunction.

Aortic Insufficiency (Aortic Regurgitation)

* Valve incompetence causing blood to leak back into the left ventricle.
* Leads to volume overload and heart failure.

Carnitine Deficiency

* Metabolic disorder affecting fatty acid oxidation.
* Can cause cardiomyopathy, muscle weakness, and hypoglycemia.

Coccidioidomycosis and Valley Fever

* Fungal infection endemic in the southwestern US.
* Can cause pulmonary symptoms and disseminated disease, rarely involving the heart.

Congenital Mitral Stenosis

* Narrowing of the mitral valve from birth.
* Causes left atrial enlargement, pulmonary hypertension, and heart failure.

Dilated Cardiomyopathy (DCM)

* Enlargement and impaired contraction of the ventricles.
* Causes heart failure, arrhythmias, and thromboembolism.

Glomerulonephritis

* Inflammation of the kidney glomeruli.
* Can cause acute kidney injury, hematuria, and proteinuria.

Heart Failure

* Clinical syndrome from structural or functional cardiac disorder impairing ventricular filling or ejection.
* Symptoms include dyspnea, fatigue, and fluid retention.

Histoplasmosis

* Fungal infection from inhaled spores.
* Usually pulmonary but can disseminate, rarely affecting the heart

Kawasaki Disease (KD)

* Acute systemic vasculitis in children (1–5 years old).
* Acute cardiac complications:
  + Myocarditis causes heart failure and arrhythmias.
  + Coronary artery aneurysms risking thrombosis, rupture, or myocardial infarction.
  + Pericarditis and valvular abnormalities.
* Prompt treatment with IV immunoglobulin and aspirin reduces complications.

Rash (in context of KD and other systemic diseases)

* Common in systemic vasculitis and infections.
* Helps in clinical diagnosis.

Pediatric Aortic Regurgitation / Mitral Regurgitation / Valve Insufficiencies

* Valve incompetence causing volume overload and heart failure.
* May be congenital or acquired (e.g., from infection or inflammation).

Pediatric Bacterial Endocarditis

* Infection of heart valves.
* Causes valve destruction, insufficiency, and systemic emboli.

Pediatric Cardiac Tumors

* Rare; may cause obstruction, arrhythmias, or heart failure.

Pediatric Dilated Cardiomyopathy

* Similar to adult DCM, it causes heart failure in children.

Pediatric HIV Infection

* Can cause cardiomyopathy, opportunistic infections, and systemic illness.

Pediatric Infective Pericarditis and Malignant Pericardial Effusion

* Infection or malignancy causing pericardial inflammation and fluid accumulation.
* Can lead to tamponade and hemodynamic compromise.

Pediatric Mitral Valve Prolapse

* Abnormal valve leaflet motion.
* Usually benign but can cause regurgitation.

Pediatric Non Viral and Viral Myocarditis

* Inflammation of myocardium from infectious or autoimmune causes.
* Can cause acute heart failure and arrhythmias.

Pediatric Septic Arthritis

* Infection of joint space.
* Not cardiac but an important differential in febrile children.

Pediatric Valvar Aortic Stenosis

* Narrowing of the aortic valve causing obstruction.
* Leads to left ventricular hypertrophy and heart failure.

Systemic Lupus Erythematosus (SLE)

* Autoimmune disease with multisystem involvement.
* Can cause pericarditis, myocarditis, endocarditis, and vasculitis.

Transient Synovitis

* Self-limited hip inflammation in children.
* Important to differentiate from septic arthritis.

**Recent guidelines or updates**

Rheumatic fever (RF) and Rheumatic heart disease (RHD) are a preventable public health problem in low- and middle-income countries and in marginalized communities in middle- and high-income countries. RF is an autoimmune inflammatory reaction to throat infections (pharyngitis) or possibly to superficial skin and skin structure infections caused by Streptococcus pyogenes, a group A beta‐hemolytic Streptococcus (GAS) bacterium. The first episode of RF is commonly seen in children aged 5 to 14 years. Recurrent episodes are most common within 1 year of the first episode but can occur throughout the life course. RHD is characterized by chronic structural and/or functional changes in the heart, most commonly in the valves, caused by one or more episodes of rheumatic fever (RF). RHD most commonly starts in childhood with a diagnostic peak in young adults aged 20 to 39 years. RHD can lead to death or lifelong disability, however, effective early intervention can prevent premature morbidity and mortality.

The WHO guideline on the prevention and diagnosis of rheumatic fever (RF) and rheumatic heart disease (RHD) provides evidence-informed recommendations for the prevention and management of RF and RHD. It encompasses three areas; 1) primary prevention of rheumatic fever and rheumatic heart disease, specifically the identification and treatment of suspected group A (beta-hemolytic) *Streptococcus* (GAS) pharyngitis and skin infections; 2) secondary prevention of recurrent rheumatic fever and of rheumatic heart disease, specifically use of long-term antibiotic prophylaxis, interventions to increase adherence to antibiotic prophylaxis, and screening for early rheumatic heart disease; and 3) management of rheumatic fever, specifically the treatment with anti-inflammatory drugs.

The recommendations are intended for a wide audience involved in the prevention and management of RF and RHD.

**Epidemiology data**

Epidemiology:

1. The global burden of RHD continues to be significant. In 2017, there were an estimated 38-40.8 million cases of RHD globally, with the highest prevalence in Oceania, South Asia, and sub-Saharan Africa.
2. Prevalence ranged from 3.4 cases/100,000 in non-endemic regions, to >1,000/100,000 cases in endemic areas.
3. Data on RHD related morbidity and mortality are less robust, but the estimate is at least 260,000-300,000 deaths per year.
4. A global registry of 3,300 RHD cases from 14 lower- to mid-income countries reveals that most patients with RHD are young (median age 28 years), female (66%), with moderate to severe multivalvular disease (64%) complicated by congestive heart failure (33%), pulmonary hypertension (29%), atrial fibrillation (22%), and stroke (7%).
5. The World Heart Federation (WHF) has set forth an aim to reduce the burden of RHD by 25% in 2025.

Rheumatic heart disease (RHD) is a major public health problem in resource-poor countries, especially in sub-Saharan Africa where about 1 million affected children between 5 and 14 years of age live with the disease. RHD still remains a major cardiovascular health problem in Nigeria. There is therefore a need for more recent clinical studies on the contemporary pattern of RHD in Nigeria. In addition, community based screening for RHD is needed to determine the true burden of the disease in Nigeria. Finally, primary and secondary preventive measures are needed to help reduce the burden of RHD in Nigeria.

It is estimated that 33 million people have RHD worldwide. Furthermore, approximately 319,400 individuals died from RHD in 2015, while about 471,000 new cases were diagnosed annually. RHD is also the most common cause of cardiovascular disease (CVD) in young people aged 25 years or below, majority of whom live in Africa, the South Pacific, Middle East, Central, and South Asia. It afflicts them in their most productive years with associated high mortality and high disability-adjusted life years. It is also a major contributor to maternal and perinatal mortality in the aforementioned regions. RHD was once common in high income countries, however by the 1980s, the condition had been almost eliminated in these countries.

The lesson from high-income countries and some developing countries is that the disease can be controlled through comprehensive, register-based RHD control programs.In Nigeria, the true burden of the disease is not known, and there is limited population based data for informed decision-making. According to WHO, the country belongs to the group of countries where it is estimated that between 1000 and 4999 people died from the disease in 2002.

in Eastern Nigeria and 1956 by Beet in Northern Nigeria. Beets examined all cardiac cases admitted to three main hospitals in Northern Nigeria (Kano, Kaduna, and Jos) and reported that RHD was responsible for 23% of cardiac conditions. Probably due to few data from Southern Nigeria at that time, he assumed that the high burden was only peculiar to Northern Nigeria and might not be applicable to coastal areas of West Africa. However, subsequent work in Ibadan by Lauckner *et al.* using an analysis of medical admissions to the University College Hospital Ibadan in 1958,showed that acute carditis was responsible for about 18.7% of all cardiac admissions. Lauckner *et al.,* therefore, concluded that with further experience as well as improved diagnostic facilities at the hospital, RHD was by no means uncommon in Nigeria

RHD is mainly a disease that begins in childhood. It has its peak incidence in children aged 5–15 years and young adults below the age of 30 years. Affected children in the absence of any intervention, progress to severe disease and eventually die before they reach adulthood. However, where the disease is milder or adequate care is available, affected children live with the disease carrying it into adulthood.

The general pattern of RHD in Nigeria is similar to the established pattern in many tropical and subtropical climates It is highly prevalent in the low socioeconomic class of the population with an infrequent and atypical nature of preceding history of RF in patients with established RHD. It is also associated with an early onset of established rheumatic valve disease and pulmonary hypertension, rapid progression of the disease, and frequent presentation as HF, especially in young adults. Furthermore, the comparatively low frequency of pure mitral stenosis (MS), the predominance of pure mitral regurgitation (MR) and mixed mitral valve lesions and a relatively low frequency of atrial fibrillation (AF) despite the severity of the mitral valve lesion was highlighted

In one report, majority (71%) of the patients were in the 6–10 year age group, 10% were aged 3–5 years while those aged 11–15 years constituted 19%. In general, Nigerian children are susceptible to RHD between the ages of 3 and 15 years In many mixed studies, the mean age at presentation ranges from 26.6 to 27.6. years (range 12–70 years), In a recent review of adult RHD cases in the Abeokuta Heart Disease Registry, a higher mean age of 43 years was reported. shows age distribution of cases in the Abeokuta Heart Disease Registry.

Socioeconomic class

Poverty and social disadvantage are important drivers and predisposing factors to the RHD scourge globally. Poverty and social disadvantage drive overcrowding, poor ventilation, malnutrition, poor sanitation and hygiene, and the poor access to healthcare associated with the development of RHD in Nigeria and elsewhere. This was classically demonstrated by Cole *et al.* in Ibadan where majority of their patients though living in urban and peri-urban areas of Ibadan, were from low socioeconomic quintiles. Similarly, the report by Jaiyesimi and Antia in 1981 on children from Ibadan showed that most of the children (90%) with RHD came from poor homes which were most often over-crowded. The number of siblings per patient ranged from 2 to 14, with a mean of 5. Recent reports still show the predominance of poverty and poverty related factors in families of patients with RHD in Nigeria.

RHD occurs after an episode or recurrent episodes of untreated or poorly treated acute RF. A past history of RF though relevant may not be readily volunteered or even remembered by many patients, especially adults with RHD. In addition, many people, especially children and adolescents affected with RHD present with severe HF as a result of severe valvular insufficiency resulting from chronic inflammation with deformed and fibrosed valvular apparatus, while the adults present with AF and embolic phenomenon in addition to the HF.

The common symptoms at presentation include cough (87.9%), easy fatigability (50.5), dyspnea on exertion (100%), orthopnea (100%), and paroxysmal nocturnal dyspnea (74.8%). Other symptoms include leg swelling (74.8%), palpitation (33.6%), chest pain (50.5%), hemoptysis (16.8%), and fever (7.5%). In terms of clinical signs, elevated jugular venous pressure (JVP), basal crepitations, displaced apex beat, and third heart sounds were common, occurring in 66.4%, 77.6%, 39.3%, 174.3%, and 5.8%, respectively. Other signs include diastolic murmur (8.4%), hepatomegaly (74.8), splenomegaly (8.4%), and ascites (33.6%).

28.3% of the patients presented in pregnancy. In an early study of HF and pregnancy at the Ahmadu Bello University Teaching Hospital, Kaduna, North Western Nigeria and Abengowe. reported that RHD was responsible for 30.6% of cases, 53.3% presented in the postpartum period while the rest antepartum Cole and Adeleye at Ibadan studied 44 pregnancies in 32 Nigerian women between the ages of 16 and 38 years (mean 27 years) with RHD. Majority of the women were gravida three, but were closely followed by the primigravidae. Pure MR was the most common valve lesion encountered. Acute pulmonary edema was common, occurring in 11 patients and which was responsible for 2 of the 3 maternal deaths. Congestive cardiac failure was the second commonest presentation (in 7) and infective endocarditis occurred in 4 patients. The third maternal death was in a woman with mitral valve prosthesis who was also on warfarin. The major precipitating factors for HF include infective endocarditis, AF with uncontrolled ventricular rate, anemia, and chest infection. In the series reported by Abengowe, congestive HF was more common than acute pulmonary edema. The perinatal mortality was 11/1000 and average birth weight was 2.7 kg, but there were five cases of low-birth-weight babies. At 6 months' postpartum, there was evidence of progression in eight

About 20% of children with RHD die within 6 years of onset of the illness. About half of the deaths occurred within the 1st year. Most deaths occurred as a result of HF or bacterial endocarditis.

A poor socioeconomic status, delays in seeking medical care, and recurrence of the RF were associated with poor prognosis, as were the presence of MS, multiple valve lesions, presence of elevated pulmonary pressure, bacterial endocarditis, and pulmonary embolism. Outcome was not affected by age, gender and PR-interval.

The authors concluded that in “developing countries where primary prevention of RF might be vitiated by poverty, ignorance and scarcity of health facilities, the prognosis in childhood RHD can be improved by simple and relatively inexpensive measures like health education, effective SBE prophylaxis and prevention of recurrent ARF. Mitral valvotom, where feasible, can improve not only the chances of survival of but also the quality of life in children with disabling mitral stenosis.

Recently screened 4107 children (mean age of 11.3 years), 53.7% females in urban and peri urban areas of Lagos. They detected 38 children with abnormal echocardiograms, of which 11 (0.27%) showed RHD including two cases of definite RHD giving a prevalence of 2.7/1000 (2.9/1000 in the peri urban, 2.4/1000 in the urban area). Echocardiography was reported to detected RHD 10 times better than auscultation (echocardiography 11 (0.27%) vs. auscultation 1 (0.02%); *P* = 0.003]. The remaining 27 children with abnormal echocardiograms had congenital heart defects (CHD) giving a prevalence of 6.6/1000 for CHD, a yield higher than for RHD.

REFERENCES

<https://www.who.int/news-room/fact-sheets/detail/rheumatic-heart-disease>

[Rheumatic Heart Disease: Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/21485-rheumatic-heart-disease)

[Medscape Registration](https://emedicine.medscape.com/article/891897-differential?form=fpf)

[Nigerian Journal of Cardiology](https://journals.lww.com/nijc/fulltext/2020/17010/rheumatic_heart_disease_in_nigeria__a_review.4.aspx)

Ogah, Okechukwu S.1,; Bode-Thomas, Fidelia2; Yilgwan, Christopher2; Ige, Olukemi2; Ogah, Fisayo3; Ogunkunle, Oluwatoyin O4; Okwuonu, Chimezie5; Sani, Mahmoud6

[Author Information](https://journals.lww.com/nijc/fulltext/2020/17010/rheumatic_heart_disease_in_nigeria__a_review.4.aspx)

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**Hypertensive Heart Failure**

**Definition and description**

Heart failure refers to a clinical syndrome where the heart cannot pump sufficiently to meet the body’s needs. It can result from various etiologies, including coronary artery disease, valvular dysfunction, and indeed hypertensive heart disease.

**Heart failure means that the heart is unable to pump blood around the body properly. It usually happens because the heart has become too weak or stiff.**

It's sometimes called congestive heart failure, although this name is not widely used now.

Heart failure does not mean your heart has stopped working. It means it needs some support to help it work better.

It can occur at any age, but is most common in older people.

Heart failure is a long-term condition that tends to get gradually worse over time.

It cannot usually be cured, but the symptoms can often be controlled for many years.

**Causes**

Heart failure is often the result of a number of problems affecting the heart at the same time.

Conditions that can lead to heart failure include:

* coronary heart disease – where the arteries that supply blood to the heart become clogged up with fatty substances (atherosclerosis), which may cause angina or a heart attack
* high blood pressure – this can put extra strain on the heart, which over time can lead to heart failure
* conditions affecting the heart muscle (cardiomyopathy)
* heart rhythm problems (arrhythmias), such as atrial fibrillation
* damage or other problems with the heart valves
* congenital heart disease – birth defects that affect the normal workings of the heart

Sometimes obesity, anemia, drinking too much alcohol, an overactive thyroid or high pressure in the lungs (pulmonary hypertension) can also lead to heart failure.

**Risk factors**

Diseases and conditions that increase the risk of heart failure include:

* **Coronary artery disease.** Narrowed arteries may limit the heart's supply of oxygen-rich blood, resulting in weakened heart muscle.
* **Heart attack.** A heart attack is a form of coronary artery disease that occurs suddenly. Damage to the heart muscle from a heart attack may mean the heart can no longer pump as well as it should.
* **Heart valve disease.** Having a heart valve that doesn't work properly raises the risk of heart failure.
* **High blood pressure.** The heart works harder than it has to when blood pressure is high.
* **Irregular heartbeats.** Irregular heartbeats, especially if they are very frequent and fast, can weaken the heart muscle and cause heart failure.
* **Congenital heart disease.** Some people who develop heart failure were born with changes in the structure or function of their heart.
* **Diabetes.** Having diabetes increases the risk of high blood pressure and coronary artery disease.
* **Sleep apnea.** This inability to breathe properly during sleep results in low blood-oxygen levels and an increased risk of irregular heartbeats. These things can lead to a weakened heart.
* **Obesity.** People who have obesity have a higher risk of developing heart failure.
* **Viral infections.** Some viral infections can damage the heart muscle.

Medicines that may increase the risk of heart failure include:

* **Some diabetes medicines.** The diabetes medicines rosiglitazone (Avandia) and pioglitazone (Actos) have been found to increase the risk of heart failure in some people. Don't stop taking these medicines without first talking to your healthcare professional.
* **Some other medicines.** Other medicines that may lead to heart failure or heart conditions include nonsteroidal anti-inflammatory drugs (NSAIDs) and some medicines used to treat high blood pressure, cancer, blood conditions, irregular heartbeats, nervous system diseases, mental health conditions, lung and urinary conditions, and infections.

Other risk factors for heart failure include:

* **Aging.** The heart's ability to work decreases with age, even in healthy people.
* **Alcohol use.** Drinking too much alcohol may weaken the heart muscle and lead to heart failure.
* **Smoking or using tobacco.** If you smoke, quit. Using tobacco increases the risk of heart disease and heart failure.

**Symptoms**

The main symptoms of heart failure are:

* breathlessness after activity or at rest
* feeling tired most of the time and finding exercise exhausting
* feeling lightheaded or fainting
* swollen ankles and legs

Some people also experience other symptoms, such as a persistent cough, a fast heart rate and dizziness.

Symptoms can develop quickly (acute heart failure) or gradually over weeks or months (chronic heart failure).

**Diagnosis methods**

**Stages of heart failure**

When you're diagnosed with heart failure, your doctor will usually be able to tell you what stage it is.

The stage describes how severe your heart failure is.

It's usually given as a class from 1 to 4, with 1 being the least severe and 4 being the most severe:

* class 1 – you don't have any symptoms during normal physical activity
* class 2 – you're comfortable at rest, but normal physical activity triggers symptoms
* class 3 – you're comfortable at rest, but minor physical activity triggers symptoms
* class 4 – you're unable to carry out any physical activity without discomfort and may have symptoms even when resting

Knowing the stage of your heart failure will help your doctors decide which treatments they think are best for you.

**Treatments**

Treatment for heart failure usually aims to control the symptoms for as long as possible and slow down the progression of the condition.

How you're treated will depend on what is causing your heart failure.

Common treatments include:

* lifestyle changes – including eating a healthy diet, exercising regularly and stopping smoking
* medicine – a range of medicines can help; many people need to take 2 or 3 different types
* devices implanted in your chest – these can help control your heart rhythm
* surgery – such as a bypass operation or a heart transplant

Treatment will usually be needed for life.

A cure may be possible when heart failure has a treatable cause. For example, if your heart valves are damaged, replacing or repairing them may cure the condition.

**For most people, heart failure is a long-term condition that can't be cured. But treatment can help keep the symptoms under control, possibly for many years.**

The main treatments are:

* healthy lifestyle changes
* medication
* devices implanted in your chest to control your heart rhythm
* surgery

In many cases, a combination of treatments will be required.

Treatment will usually need to continue for the rest of your life.

**Care plan**

If you have heart failure, you and everyone involved in your care will be given a care plan.

This should include:

* plans for managing your heart failure, including follow-up care, rehabilitation and access to social care
* symptoms to look out for in case your condition worsens
* details of how to contact your care team or specialist

The care plan should be reviewed at least every 6 months by your GP.

**Lifestyle changes**

Having a healthy lifestyle, including eating a balanced diet, doing exercise and not smoking, can help with your symptoms and reduce your risk of becoming seriously ill.

You should be offered an exercise-based cardiac rehabilitation program.

**Medicines for heart failure**

Most people with heart failure are treated with medication. Often you'll need to take 2 or 3 different medicines.

Some of the main medicines for heart failure include:

* ACE inhibitors
* angiotensin-2 receptor blockers (ARBs or AIIRAs)
* beta blockers
* mineralocorticoid receptor antagonists
* diuretics
* ivabradine
* sacubitril valsartan
* hydralazine with nitrate
* digoxin
* SGLT2 inhibitors

You may need to try a few different medicines before you find a combination that controls your symptoms but doesn't cause unpleasant side effects.

**ACE inhibitors**

Angiotensin-converting enzyme (ACE) inhibitors work by relaxing and opening up your blood vessels, which makes it easier for your heart to pump blood around the body.

Examples of ACE inhibitors include ramipril, captopril, enalapril, lisinopril and perindopril.

The most common side effect of ACE inhibitors is a dry, irritating cough.

If you have a troublesome cough, an ACE inhibitor may be switched to an ARB.

ACE inhibitors can also cause your blood pressure to fall too low, and they may cause kidney problems. Your GP will monitor this.

**Angiotensin-2 receptor blockers (ARBs)**

Angiotensin-2 receptor blockers (ARBs) work in a similar way to ACE inhibitors by relaxing blood vessels and reducing blood pressure.

They tend to be used as an alternative to ACE inhibitors because they don't usually cause a cough, although they may not be quite as effective as ACE inhibitors.

Examples of ARBs include candesartan, losartan, telmisartan and valsartan.

Side effects of ARBs can include low blood pressure and high levels of potassium in your blood.

Your doctor will carry out regular blood tests to monitor your potassium level.

**Beta blockers**

Beta blockers work by slowing your heart down and protecting your heart from the effects of adrenaline and noradrenaline, "fight or flight" chemicals produced by the body.

There are several different beta blockers, but the main ones used to treat heart failure in the UK are bisoprolol, carvedilol and nebivolol.

Possible side effects include dizziness, tiredness and blurred vision.

But most people taking them have either no or very mild side effects that become less troublesome with time.

**Mineralocorticoid receptor antagonists (MRAs)**

MRAs make you pass more urine, and help lower blood pressure and reduce fluid around the heart, but they don't reduce potassium levels.

The most widely used MRAs are spironolactone and eplerenone.

Spironolactone may cause enlarged breasts in men (gynaecomastia) and breast tenderness and increased hair growth in women.

Eplerenone can cause sleeping difficulties, dizziness and headaches.

The most serious side effect of these medicines is that they can cause the level of potassium in your blood to become dangerously high.

Your doctor will carry out regular blood tests to check for this.

**Diuretics**

Diuretics (water pills) make you pass more urine and help relieve ankle swelling and breathlessness caused by heart failure.

There are many different types of diuretic, but the most widely used for heart failure are furosemide (also called frusemide) and bumetanide.

Possible side effects of diuretics include dehydration and reduced levels of sodium and potassium in the blood.

**Ivabradine**

Ivabradine is a medicine that can help slow your heart down.

It's a useful alternative to beta blockers if you can't take them or they cause troublesome side effects.

It can also be used alongside beta blockers if they don't slow the heart enough.

Possible side effects include headaches, dizziness and blurred vision.

**Sacubitril valsartan**

Sacubitril valsartan is a single tablet that combines an ARB and a medication called a neprilysin inhibitor.

It's suitable for people with more severe heart failure, whose heart is only able to pump a reduced amount of oxygenated blood around the body despite taking other medication.

The most common side effects of sacubitril valsartan are low blood pressure, high potassium levels and kidney problems.

**Hydralazine with nitrate**

Hydralazine in combination with nitrate can help relax and open up the blood vessels.

These medicines are sometimes prescribed by heart specialists (cardiologists) for people who are unable to take an ACE inhibitor or ARB.

Side effects can include headaches, a fast heartbeat and a pounding, fluttering or irregular heartbeat (palpitations).

**Digoxin**

Digoxin can improve your symptoms by strengthening your heart muscle contractions and slowing down your heart rate.

It's normally only recommended for people who have symptoms despite treatment with ACE inhibitors, ARBs, beta blockers and diuretics.

Possible side effects include dizziness, blurred vision, feeling and being sick, diarrhoea and an irregular heartbeat.

**SGLT2 inhibitors**

SGLT2 inhibitors are tablets that can help lower your blood sugar levels.

Empagliflozin and dapagliflozin are types of SGLT2 inhibitor. They can be used to treat some types of heart failure, as an add-on to other medicines.

Possible side effects include thrush, peeing more than usual, a mild skin rash and back pain.

**Take your medication**

It's very important that you take any prescribed medication, even if you begin to feel better.

Check with your care team if:

* other medicines might interfere with your medication
* you experience any side effects

**Devices for heart failure**

Some people with heart failure will need to have a procedure to implant a small device in their chest that can help control or monitor their heart's rhythm.

The most commonly used devices are:

* pacemakers
* cardiac resynchronization therapy (CRT) devices
* implantable cardioverter defibrillators (ICDs)
* CRT-Ds

**Pacemakers**

You may need to have a pacemaker fitted if your heart beats too slowly.

A pacemaker monitors your heart rate continuously, and sends electrical pulses to your heart to keep it beating regularly and at the right speed.

The pacemaker is implanted under the skin by a cardiologist, usually under local anesthetic.

You'll usually need to stay in the hospital overnight to check it's working properly. Serious complications are unusual.

**Cardiac resynchronization therapy**

In some people with heart failure, the walls of the main pumping chamber (the left ventricle) don't work together and contract out of sync with each other.

Cardiac resynchronization therapy (CRT) is a special type of pacemaker that can correct the problem by making the walls of the left ventricle all contract at the same time. This makes the heart pump more efficiently.

Most pacemakers only have 1 or 2 wires to the heart, but CRT requires an extra wire.

**Implantable cardioverter defibrillators (ICDs)**

People who have, or are at high risk of developing, an abnormal heart rhythm may need to have a device known as an implantable cardioverter defibrillator (ICD) fitted.

An ICD constantly monitors the heart rhythm.

If the heart starts beating dangerously fast, the ICD will try to bring it back to normal by giving it a small, controlled electrical shock (defibrillation).

If this fails, the ICD will deliver a larger shock.

As with pacemakers, ICDs are implanted in hospitals, usually under local anesthetic.

Like pacemakers, you'll need to avoid things that can interfere with the way the ICD works, such as airport security systems.

**CRT-Ds**

Devices that combine cardiac resynchronization and defibrillation are implanted into patients who need both.

These combination devices are usually called CRT-Ds.

**Pulmonary artery pressure sensors**

Some people with chronic heart failure may need to have a device known as a pulmonary artery pressure sensor fitted.

It's implanted into your artery in hospital, under local anesthetic.

The sensor sends blood pressure measurements to a monitor in your home. The monitor sends the measurements to your care team, to help them decide whether your treatment needs to be changed. This should help to manage your treatment and reduce the chance of you being admitted to hospital.

This is a new procedure that might not yet be available to everyone.

**Improving muscle strength**

If you are having a bad flare-up and are unable to exercise, you may be offered electrical stimulation to make your muscles stronger.

This is where electrodes are placed on your skin and small electrical impulses are sent to weak muscles, usually in your arms or legs.

**Surgery**

Medicines are the main treatment for heart failure, but for some people surgery may help.

Operations that can help with heart failure include:

* heart valve surgery
* a coronary angioplasty or bypass
* left ventricular assist devices
* heart transplant

**Heart valve surgery**

If the valves of your heart are damaged or diseased, your doctor may suggest valve surgery.

There are 2 types of valve surgery: valve replacement and valve repair.

The type of surgery you have will depend on what's wrong with the valve and how serious the problem is.

Your doctor will discuss this with you.

**Angioplasty or bypass**

If your heart failure is related to coronary heart disease, your doctor may recommend a:

* coronary angioplasty – where a tiny balloon is used to stretch open a narrowed or blocked artery; usually a small mesh tube called a stent is put in the artery to support it
* coronary artery bypass graft (CABG) – where a blood vessel from another part of the body is used to divert blood around narrowed or clogged parts of an artery

These procedures will help make it easier for your heart to pump blood around your body.

**Left ventricular assist devices**

Left ventricular assist devices (LVADs) are mechanical pumps that can help if your left ventricle isn't working properly and medication alone isn't helping.

They may be used as a permanent treatment if you can't have a heart transplant, or as a temporary measure while you wait for a transplant.

In addition to the pump, LVADs also include an external battery. A wire connecting this to the pump will need to be placed under your skin during the operation.

**Heart transplant**

A heart transplant may be necessary if you develop severe heart failure that can't be treated effectively with medication or other types of surgery.

A heart transplant is a complex procedure that carries serious risks, so it's not suitable for everyone with severe heart failure.

There's also a shortage of hearts for transplantation, so some people have to wait years for a suitable donor heart to become available.

**Prevention**

One way to prevent heart failure is to treat and control the conditions that can cause it. These conditions include coronary artery disease, high blood pressure, diabetes and obesity.

Some of the same lifestyle changes used to manage heart failure also may help prevent it. Try these heart-healthy tips:

* Don't smoke.
* Get plenty of exercise.
* Eat healthy foods.
* Maintain a healthy weight.
* Reduce and manage stress.
* Take medicines as directed.

**Prognosis**

**Outlook for heart failure**

Heart failure is a serious long-term condition that will usually continue to get slowly worse over time.

It can severely limit the activities you're able to do and is often eventually fatal.

But it's very difficult to tell how the condition will progress on an individual basis.

It's very unpredictable. Lots of people remain stable for many years, while in some cases it may get worse quickly.

**Complications**

If you have heart failure, it's important to have regular health checkups, even if symptoms improve. Your healthcare professional can examine you and run tests to check for complications.

Complications of heart failure depend on your age, overall health and the severity of heart disease. They may include:

* **Kidney damage or failure.** Heart failure can reduce the blood flow to the kidneys. Untreated, this can cause kidney failure. Kidney damage from heart failure can require dialysis for treatment.
* **Other heart changes.** Heart failure can cause changes in the heart's size and function. These changes may damage heart valves and cause irregular heartbeats.
* **Liver damage.** Heart failure can cause fluid buildup that puts too much pressure on the liver. This fluid backup can lead to scarring, which makes it more difficult for the liver to work properly.
* **Sudden cardiac death.** If the heart is weak, there is a risk of dying suddenly due to a dangerous irregular heart rhythm.

**Dealing with medicine side effects**

If you are given medicine to manage your heart failure, your doctor or specialist heart failure nurse will start you on a low dose and increase it until they get the best results for you.

However, like all medicines, they can cause side effects, such as feeling lightheaded and dizzy.

If you experience any side effects, do not just stop taking your medicines. Instead, discuss these side effects with your nurse, GP or cardiologist. They may suggest taking a lower dose or taking a different medicine.

Getting the right balance and dose of your medicine is important so you can feel as well as possible. However, be aware that this can take time.

Diuretics work differently to other heart failure medicines. They reduce the build-up of fluid in your body, which happens when your heart is not working properly, by helping the kidneys get rid of excess water in your pee.

Normally you will be advised to take your diuretics at a time that means you do not have to get up too often in the night.

If you have a day out planned, think about the best time to take them so you do not need to pee as often, and check with your nurse or doctor that they are happy with the plan.

**When to see a doctor**

See your healthcare professional if you think you might have symptoms of heart failure. Call emergency medical help if you have any of the following:

* Chest pain.
* Fainting or severe weakness.
* Rapid or irregular heartbeat with shortness of breath, chest pain or fainting.
* Sudden, severe shortness of breath and coughing up white or pink, foamy mucus.

These symptoms may be due to heart failure. But there are many other possible causes. Don't try to diagnose yourself.

At the emergency room, healthcare professionals do tests to learn if your symptoms are due to heart failure or something else.

Call your healthcare professional right away if you have heart failure and:

* Your symptoms suddenly get worse.
* You develop a new symptom.
* You gain 5 pounds (2.3 kilograms) or more within a few days.

Such changes could mean that existing heart failure is getting worse or that treatment isn't working.

**Differential diagnosis**

**Chronic Obstructive Pulmonary Disease (COPD)**

Chronic obstructive pulmonary disease (COPD) is a group of progressive lung diseases, including emphysema and chronic bronchitis, that cause difficulty in breathing due to airway obstruction. COPD and heart failure share common symptoms such as shortness of breath, fatigue, and swelling in the legs, making it a key differential diagnosis to consider.

**Key Features of COPD:**

Dyspnea (shortness of breath) is the hallmark symptom, often triggered by physical activity or respiratory infections.

Cough and sputum production are common, especially in patients with chronic bronchitis.

Patients with COPD may have a history of smoking or exposure to other lung irritants.

**Distinguishing COPD from Heart Failure:**

In heart failure, dyspnea typically occurs at rest or with minimal exertion and may worsen when lying flat (orthopnea) or during the night (paroxysmal nocturnal dyspnea).

Chest X-rays and pulmonary function tests can help differentiate the two conditions. In COPD, chest X-rays may show hyperinflation, and pulmonary function tests will show obstructive patterns, while heart failure often reveals signs of fluid retention in the lungs.

Echocardiograms can assess heart function and identify left or right ventricular dysfunction, which is characteristic of heart failure but absent in COPD.

**Treatment Overlap:**

Both conditions benefit from bronchodilators, oxygen therapy, and diuretics. However, heart failure treatment typically involves medications like ACE inhibitors, beta-blockers, and mineralocorticoid antagonists, which are not commonly used in COPD.

**2. Pulmonary Embolism (PE)**

Pulmonary embolism (PE) occurs when a blood clot travels to the lungs, blocking a pulmonary artery and reducing blood flow to lung tissue. This can lead to symptoms similar to those of heart failure, such as chest pain, shortness of breath, and dizziness.

**Key Features of Pulmonary Embolism:**

Sudden onset of symptoms, often including sharp, pleuritic chest pain, which is different from the more gradual onset of heart failure symptoms.

Hemoptysis (coughing up blood) may be present in some cases.

Risk factors include recent surgery, prolonged immobility, deep vein thrombosis (DVT), or a history of clotting disorders.

**Distinguishing PE from Heart Failure:**

Clinical history plays a significant role. A recent history of surgery, immobility, or DVT makes PE more likely.

CT pulmonary angiography is the gold standard for diagnosing PE, while echocardiography and chest X-rays are more commonly used to diagnose heart failure.

D-dimer levels may be elevated in both conditions, but this test is non-specific. A high clinical suspicion and confirmatory imaging are essential.

**Treatment Overlap:**

Both conditions may require hospitalization and oxygen therapy.

However, PE is treated primarily with anticoagulation therapy (e.g., heparin or warfarin), whereas heart failure management involves medications like ACE inhibitors, diuretics, and beta-blockers.

**3. Acute Myocardial Infarction (AMI)**

Acute myocardial infarction (AMI), or heart attack, occurs when there is a blockage in one of the coronary arteries, resulting in damage to the heart muscle. AMI can present with symptoms such as chest pain, dyspnea, and fatigue, which may resemble heart failure symptoms, especially if heart failure develops after the infarction.

**Key Features of Acute Myocardial Infarction:**

Severe chest pain, often described as crushing or pressure-like, that may radiate to the arm, jaw, or back.

Sweating, nausea, and vomiting may accompany the pain.

Patients may experience elevated cardiac biomarkers (e.g., troponin), which help confirm the diagnosis.

**Distinguishing AMI from Heart Failure:**

In AMI, the chest pain is typically more severe and is associated with cardiac biomarkers that indicate myocardial injury.

An electrocardiogram (ECG) will show characteristic changes, such as ST-segment elevation or depression, or the presence of pathological Q waves in AMI, whereas heart failure does not typically cause such changes.

Echocardiography in heart failure may show global or regional ventricular dysfunction, but in the case of AMI, there may be a more localized area of myocardial damage.

**Treatment Overlap:**

Both conditions may involve the use of beta-blockers, antiplatelet agents, and angiotensin-converting enzyme inhibitors.

However, the immediate treatment of AMI focuses on revascularization (e.g., with thrombolytics or percutaneous coronary intervention), while heart failure treatment centers on managing symptoms and preventing disease progression.

**4. Chronic Kidney Disease (CKD)**

Chronic kidney disease (CKD) refers to the gradual loss of kidney function over time. CKD and heart failure often coexist, and patients with one condition may eventually develop the other. Symptoms like edema, fatigue, and shortness of breath can overlap, making the differentiation between the two challenging.

**Key Features of Chronic Kidney Disease:**

Edema is common in CKD due to fluid retention, often presenting in the lower extremities.

Elevated creatinine and blood urea nitrogen (BUN) levels in blood tests are hallmarks of kidney dysfunction.

Anemia is often present in CKD, contributing to fatigue and weakness.

**Distinguishing CKD from Heart Failure:**

Urine tests can identify kidney dysfunction, such as proteinuria or hematuria, which are not typically present in heart failure.

Serum creatinine and glomerular filtration rate (GFR) are key indicators of kidney function, and levels outside the normal range suggest CKD rather than heart failure.

Echocardiography may show left or right ventricular dysfunction in heart failure, but this would not be the case in CKD unless the patient has developed heart failure as a complication of their kidney disease.

**Treatment Overlap:**

Both conditions require careful management of fluid balance and blood pressure. Diuretics may be used in both, but in CKD, managing renal function through agents like ACE inhibitors or angiotensin receptor blockers is crucial.

**5. Anemia**

Anemia, particularly iron deficiency anemia or anemia of chronic disease, can lead to symptoms like fatigue, weakness, and shortness of breath, which are similar to those seen in heart failure.

**Key Features of Anemia:**

Fatigue, paleness, and dizziness are common symptoms.

In severe cases, tachycardia (increased heart rate) and hypotension may occur.

Anemia is diagnosed through a complete blood count (CBC), which shows low hemoglobin or hematocrit levels.

**Distinguishing Anemia from Heart Failure:**

Blood tests are critical for diagnosing anemia, with low hemoglobin levels indicating the presence of anemia.

In heart failure, the ejection fraction is often reduced, while in anemia, this is typically not the case.

The presence of iron deficiency or vitamin B12 deficiency can provide further clues as to whether anemia is the primary issue.

**Treatment Overlap:**

Both conditions may involve the use of medications to improve oxygen delivery to tissues. For anemia, iron supplements or erythropoiesis-stimulating agents may be used, whereas heart failure treatment centers around improving cardiac output and fluid balance.

**Epidemiology data (e.g., how common it is, affected demographics).**

Medication adherence was significantly higher among the controls than the HHF cases with about two-thirds of the controls having high medication adherence compared to one-fifth of the HHF cases. The controls were more likely to have been on ACE-Is/ARBs, calcium channel blockers and diuretics than individuals who developed HHF.

HHF cases had higher respiratory rates than controls, but lower systolic and diastolic blood pressures than the controls. Though, there was no significant difference in pulse rate between the two groups. Significant proteinuria by dipstick (more than trace proteinuria) was observed in almost half of the HHF group but only in about one-tenth of the controls. Individuals with HHF also had significantly higher serum urea and creatinine and significantly lower estimated glomerular filtration rate (eGFR).

History of background kidney disease was associated with a 6-fold increased risk of HHF which was attenuated by 17% in age and sex adjusted analysis. Previous alcohol consumption was associated with about 4-fold increased risk of HHF which was magnified to 6-fold increased risk of HHF in age and sex adjusted analysis. Alcohol consumption was associated with 10% increased risk of HHF (crude estimate) and 13% increased risk (age and sex adjusted) per 10g or glass of alcohol consumed per day. Daily fruits and vegetables consumption were associated with 62% reduced risk of HHF which remained almost the same in age and sex adjusted analysis. Moderate medication adherence was associated with 3.88-fold increased risk of HHF which was magnified to 4.54-fold increased risk in age and sex adjusted analysis while low medication adherence was associated with about 9-fold increased risk of HHF (crude) which was magnified to about 12-fold in age and sex adjusted analysis. Use of ARBs/ACEI-s was associated with a 58% reduced risk of HHF (crude) and 64% reduced risk in age and sex adjusted estimates. Calcium channel blockers use was associated with 82% reduced risk of HHF (83% in age and sex adjusted estimates) while diuretics use was associated with 43% reduced risk of HHF (45% in age and sex adjusted estimates). Dipstick proteinuria was associated with 6-fold increased risk of HHF in both crude, and age- and sex adjusted estimates. A mg increase in serum urea was associated with 4% increased risk of HHF (both crude, and age- and sex adjusted estimates) while a mg increase in serum creatinine was associated with 3.5-fold increased risk of HHF (crude estimate) which was magnified to 4.25-fold increased risk after age and sex

**Epidemiology of Hypertensive Heart Failure in Nigeria**

Hypertensive heart failure (HHF) is a significant public health issue in Nigeria, with hypertension being the leading cause of heart failure (HF) in sub-Saharan Africa.234 In Nigeria, **hypertension accounts for up to 61% of heart failure cases in a cohort in Abuja and 75.7% in another cohort in Ibadan**. Despite advancements in hypertension care and the development of potent anti-hypertensive medications, the incidence of HHF continues to rise.

A study conducted in Ibadan, Nigeria, identified several risk factors for HHF among patients with hypertension. The study found that **low education levels (below tertiary education) and a history of kidney disease were significant risk factors for HHF**. Additionally, poor medication adherence, particularly low adherence, was strongly associated with an increased risk of HHF.

The study also highlighted the importance of calcium channel blockers in the management of hypertension and HHF. Individuals using calcium channel blockers had a significantly lower risk of developing HHF compared to those not using these medications.

Moreover, proteinuria and reduced glomerular filtration rate (GFR) were identified as significant risk factors for HHF, indicating the critical relationship between the heart and kidneys in HHF. These findings support the concept of the cardiorenal syndrome, where dysfunction in one organ system can lead to dysfunction in the other.

In summary, the epidemiology of HHF in Nigeria is characterized by a high prevalence of hypertension as the underlying cause, with significant risk factors including low education levels, history of kidney disease, poor medication adherence, and renal dysfunction.

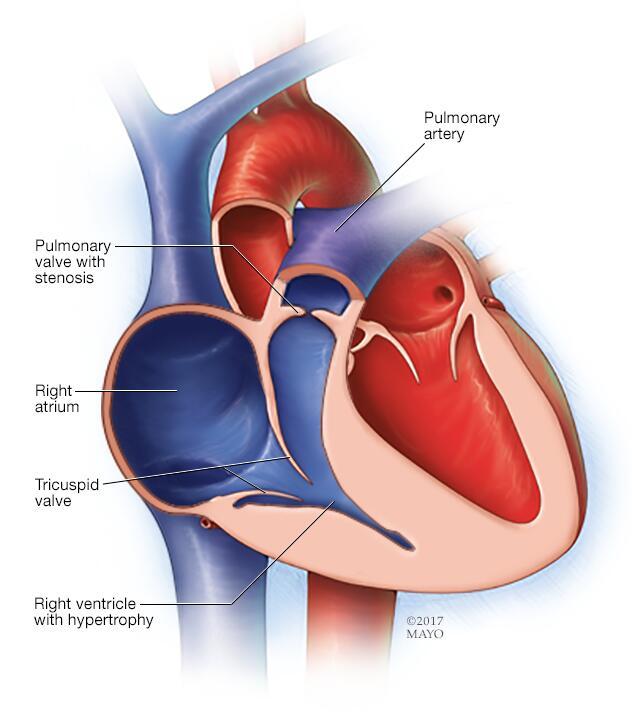
REFERENCE

[Heart failure - Living with - NHS](https://www.nhs.uk/conditions/heart-failure/living-with/)

<https://www.mayoclinic.org/diseases-conditions/heart-failure/symptoms-causes/syc-20373142>

**PULMONARY HEART STENOSIS**

Pulmonary valve stenosis is a narrowing of the valve between the lower right heart chamber and the lung arteries. In a narrowed heart valve, the valve flaps may become thick or stiff. This reduces blood flow through the valve.



### **Pulmonary stenosis**

In pulmonary stenosis, the pulmonary valve is narrowed. The greater the narrowing, the harder the lower right heart chamber, called the right ventricle, must work to pump blood to the lungs. The increased pressure causes the right ventricle to become thick.

Usually, pulmonary valve disease is caused by a heart problem that develops before birth. A heart problem present at birth is called a congenital heart defect. In adults, pulmonary valve stenosis may be a complication of another illness.

Pulmonary valve stenosis ranges from mild to severe. Some people with mild pulmonary valve stenosis don't have symptoms. They may need only occasional health checkups. Moderate and severe pulmonary valve stenosis may need a procedure to repair or replace the valve.

## 

## **Causes**

Pulmonary valve stenosis usually results from a heart problem present at birth. The exact cause is unclear. The pulmonary valve doesn't develop properly as the baby is growing in the womb.

The pulmonary valve is made of three thin pieces of tissue called flaps, also called cusps. The cusps open and close with each heartbeat. They make sure blood moves in the right direction.

In pulmonary valve stenosis, one or more of the cusps may be stiff or thick. Sometimes the cusps may be joined together. That means they are fused. So the valve doesn't open fully. The smaller opening makes it harder for blood to leave the lower right heart chamber. Pressure increases inside the chamber. The increased pressure strains the heart. Eventually the lower right heart chamber wall gets thicker.

**Risk factors**

Things that may increase the risk of pulmonary valve stenosis include:

* **German measles, also called rubella.** Having German measles during pregnancy increases the risk of pulmonary valve stenosis in the baby.
* **Noonan syndrome.** This condition is caused by altered deoxyribonucleic acid (DNA). It can lead to many problems with the heart's structure and function.
* **Rheumatic fever.** This complication of strep throat can cause permanent damage to the heart and heart valves. It increases the risk of developing pulmonary valve stenosis later in life.
* **Carcinoid syndrome.** This condition occurs when a rare cancerous tumor releases certain chemicals into the bloodstream. It causes shortness of breath, flushing and other symptoms. Some people with this syndrome develop carcinoid heart disease, which damages heart valves.

## 

## **Symptoms**

Pulmonary valve stenosis symptoms depend on how much blood flow is blocked. Some people with mild pulmonary stenosis do not have symptoms. Those with more-severe pulmonary stenosis may first notice symptoms while exercising.

Pulmonary valve stenosis symptoms may include:

* A whooshing sound called a heart murmur that can be heard with a stethoscope.
* Fatigue.
* Shortness of breath, especially during activity.
* Chest pain.
* Fainting.

Babies with pulmonary valve stenosis may have blue or gray skin due to low oxygen levels.

## **Diagnosis**

Pulmonary valve stenosis is often diagnosed in childhood. But it may not be detected until later in life.

A health care provider uses a stethoscope to listen to the heart. A whooshing sound, called a heart murmur, may be heard. The sound is caused by choppy blood flow across the narrowed valve.

Tests to diagnose pulmonary valve stenosis include:

* **Electrocardiogram (ECG or EKG).** This quick and painless test records the electrical signals in the heart. Sticky patches, called electrodes, are placed on the chest and sometimes the arms and legs. Wires connect the electrodes to a computer, which displays the test results. An ECG can show how the heart is beating and may reveal signs of heart muscle thickening.
* **Echocardiogram.** An echocardiogram uses sound waves to produce images of the heart. This common test shows how the heart beats and pumps blood. An echocardiogram can show the shape of the pulmonary valve. The test can show how much of the valve is narrowed.
* **Cardiac catheterization.** A thin tube called a catheter is inserted into the groin and threaded through the blood vessels to the heart. Dye flows through the catheter into the blood vessels to make them show up more clearly on X-rays. This part of the test is called a coronary angiogram.  
  During the test, pressures within the heart can be measured to see how forcefully blood pumps through the heart. A provider can determine the severity of pulmonary stenosis by checking the difference in pressure between the right lower heart chamber and the lung artery.
* **Other imaging tests.** Magnetic resonance imaging (MRI) and computed tomography (CT) scans are sometimes used to confirm the diagnosis of pulmonary valve stenosis.

## 

## **Treatment**

If you have mild pulmonary valve stenosis without symptoms, you may only need occasional health checkups.

If you have moderate or severe pulmonary valve stenosis, you may need a heart procedure or heart surgery. The type of procedure or surgery done depends on your overall health and the appearance of your pulmonary valve.

### **Surgeries or other procedures**

Pulmonary valve stenosis treatment may include:

* **Balloon valvuloplasty.** The provider inserts a flexible tube with a balloon on the tip into an artery, usually in the groin. X-rays help guide the tube, called a catheter, to the narrowed valve in the heart. The balloon inflates, making the valve opening larger. The balloon is deflated. The catheter and balloon are removed.  
  Valvuloplasty may improve blood flow through the heart and reduce pulmonary valve stenosis symptoms. But the valve may narrow again. Some people need valve repair or replacement in the future.
* **Pulmonary valve replacement.** If balloon valvuloplasty isn't an option, open-heart surgery or a catheter procedure may be done to replace the pulmonary valve. If there are other heart problems, the surgeon may repair those during the same surgery.  
  People who have had pulmonary valve replacement need to take antibiotics before certain dental procedures or surgeries to prevent endocarditis.

## **Self care**

If you have valve disease, it's important to take steps to keep your heart healthy. Certain lifestyle changes can decrease your risk of developing other types of heart disease or having a heart attack.

Lifestyle changes to talk about with your healthcare provider include:

* Quitting smoking.
* Eating a heart-healthy diet that includes fruits and vegetables, low-fat dairy products, whole grains, and lean meat.
* Maintaining a healthy weight.
* Getting regular exercise.

**Complications**

Possible complications of pulmonary stenosis include:

* **Infection of the lining of the heart, called infective endocarditis.** People with heart valve problems, such as pulmonary stenosis, have an increased risk of developing bacterial infections that affect the inner lining of the heart.
* **Irregular heartbeats, called arrhythmias.** People with pulmonary stenosis are more likely to have irregular heartbeats. Unless the stenosis is severe, irregular heartbeats due to pulmonary stenosis usually aren't life-threatening.
* **Thickening of the heart muscle.** In severe pulmonary stenosis, the lower right heart chamber must pump harder to force blood into the pulmonary artery. The strain on the heart causes the muscular wall of the ventricle to thicken. The condition is called right ventricular hypertrophy.
* **Heart failure.** If the right ventricle can't pump properly, heart failure eventually develops. Symptoms of heart failure include fatigue, shortness of breath, and swelling of the legs and belly area.
* **Pregnancy complications.** The risks of complications during labor and delivery are higher for those with severe pulmonary valve stenosis than for those without it.

## 

## **When to see a doctor**

Talk to your health care provider if you or your child has:

* Shortness of breath.
* Chest pain.
* Fainting.

Prompt diagnosis and treatment of pulmonary valve stenosis can help reduce the risk of complications.

**EPIDEMIOLOGY**

Isolated valvar pulmonary stenosis accounts for 7% to 12% of congenital heart diseases. Extracardiac and neurodevelopmental comorbidities affect approximately 56% of patients with pulmonary stenosis. In such cases, a molecular diagnosis is more common. For example, the *PTPN11* mutation is identified in 50% of patients with pulmonary stenosis and Noonan syndrome. Moreover, a familial form of nonsyndromic pulmonary stenosis has been described and is suspected to be related to *GATA4* mutations. Pulmonary stenosis does not seem to have any gender predilection

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of pulmonary stenosis in infants includes:

* Congenital heart defects with associated pulmonary stenosis, such as a double-chambered right ventricle, double-outlet right ventricle, absent pulmonary valve, tetralogy of Fallot, atrioventricular septal defect, atrial septal defect, and ventricular septal defect
* Pulmonary atresia with intact ventricular septum
* Ventricular septal defect

In adult patients, differential diagnoses include:

* Rheumatic valvular heart disease
* Carcinoid heart disease
* Pulmonary embolism
* Right heart failure
* Cardiac tumor
* Cardiac sarcoma

REFERENCES

[Pulmonary valve stenosis - Symptoms & causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/pulmonary-valve-stenosis/symptoms-causes/syc-20377034)

[Pulmonary valve stenosis - Diagnosis & treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/pulmonary-valve-stenosis/diagnosis-treatment/drc-20377039)

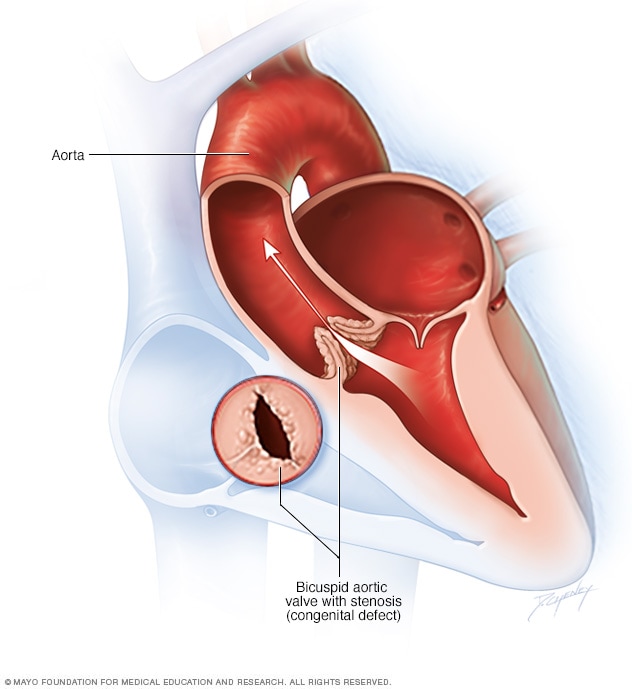
[Pulmonary Stenosis - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK560750/#article-28064.s4)

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## **Bicuspid aortic valve**

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**Bicuspid aortic valve with stenosis**

Bicuspid aortic valve is a heart condition present at birth. That means it is a congenital heart defect.

The aortic valve is between the left lower heart chamber and the body's main artery, called the aorta. Flaps of tissue on the valve open and close with each heartbeat. The flaps are called cusps. They make sure blood flows in the correct direction.

Usually the aortic valve has three cusps. A bicuspid valve has only two cusps. Rarely, some people are born with an aortic valve that has one cusp or four cusps. A valve with one cusp is called unicuspid. A valve with four cusps is called quadricuspid.

Changes to the aortic valve can cause health conditions, including:

* **Narrowing of the aortic valve, called aortic valve stenosis.** The valve may not open fully. Blood flow from the heart to the body is reduced or blocked.
* **Backward flow of blood, called aortic valve regurgitation.** Sometimes, the bicuspid aortic valve doesn't close tightly. This causes blood to flow backward.
* **Enlarged aorta, called aortopathy.** An enlarged aorta increases the risk of a tear in the lining of the aorta. This tear is called an aortic dissection.

### 

### **Symptoms**

If the bicuspid valve causes severe aortic stenosis or severe aortic regurgitation, symptoms may include:

* Chest pain.
* Shortness of breath.
* Difficulty exercising.
* Fainting or near fainting.

Most people with bicuspid aortic valve don't have symptoms of heart valve disease until they're adults. But some infants may have severe symptoms.

#### **When to see a doctor**

If you think that you or your baby has symptoms of a heart condition, make an appointment for a health checkup right away.

### **Causes**

Bicuspid aortic valve happens while the unborn baby, also called a fetus, is growing during pregnancy. Healthcare professionals aren't sure what causes most congenital heart defects, including bicuspid aortic valve. But genetics may play a role in causing bicuspid aortic valve.

### **Risk factors**

A family history of early heart disease may increase the risk of developing heart valve disease such as bicuspid aortic valve. Sometimes bicuspid aortic valve runs in families, which means it is inherited.

### **Diagnosis**

A bicuspid aortic valve may be found when tests are done for another health concern. The healthcare professional may hear a heart murmur when listening to the heart.

An echocardiogram can confirm a diagnosis of bicuspid aortic valve. This test uses sound waves to create videos of the beating heart. It shows how blood moves through the heart chambers, the heart valves and the aorta.

If you have a bicuspid aortic valve, you usually have a CT scan to check for changes in the aorta's size.

### **Treatment**

If you have a bicuspid aortic valve, you are usually sent to a doctor trained in congenital heart disease. This type of doctor is called a congenital cardiologist.

Anyone with a bicuspid aortic valve needs regular health checkups and imaging tests. Echocardiograms can check for a narrowed or leaking aortic valve. The test also looks for changes in the size of the aorta.

Treatment for a bicuspid aortic valve depends on how severe the heart valve disease is. It may include medicines or a procedure or surgery to fix or replace the valve.

#### **Medications**

There are no medicines to repair a bicuspid aortic valve. But medicines may be used to treat symptoms caused by heart valve disease. For example, your healthcare professional may recommend blood pressure medicine.

#### **Surgeries or other procedures**

Surgery may be needed if a bicuspid aortic valve is causing:

* Aortic valve stenosis.
* Aortic valve regurgitation.
* An enlarged aorta.

Surgery is done to repair or replace the aortic valve. The type of surgery done depends on the specific heart valve condition and your symptoms.

* **Aortic valve replacement.** The surgeon removes the damaged valve. It's replaced with a mechanical valve or a valve made from cow, pig or human heart tissue. The tissue valve is called a biological tissue valve. Sometimes, the aortic valve is replaced with the person's own lung valve. The lung valve is replaced with a lung tissue valve from a deceased donor. This more complicated surgery is called the Ross procedure.  
  Biological tissue valves break down over time. They may eventually need to be replaced. If you have a mechanical valve, you need to take blood thinners for life to prevent blood clots. Together, you and your healthcare team can talk about the benefits and risks of each valve type.
* **Aortic root and ascending aorta surgery.** Surgeons remove the enlarged section of the aorta located near the heart. It's replaced with a synthetic tube, called a graft, which is sewn into place. Sometimes, only the enlarged part of the aorta is removed. The aortic valve remains. The aortic valve also can be replaced or repaired during this surgery.
* **Balloon valvuloplasty.** This procedure can treat aortic valve stenosis in infants and children. In adults, the aortic valve tends to narrow again after the procedure. So it's usually only done if you're too ill for surgery or you're waiting for a valve replacement.  
  This heart valve treatment uses a thin, flexible tube called a catheter. The catheter has a balloon on the tip. The surgeon inserts the catheter into an artery in the arm or groin. Then the catheter is guided to the aortic valve. Once in place, the balloon inflates, making the valve opening bigger. The balloon is deflated. The catheter and balloon are removed.

### 

### **Lifestyle and home remedies**

Anyone born with a bicuspid aortic valve needs health checkups for life. A doctor trained in heart diseases, called a cardiologist, should examine you for changes in your condition.

People with a bicuspid aortic valve are more likely to get an infection of the inner lining of the heart's chambers and valves. This infection is called infective endocarditis. Proper dental care can help lower your risk.

A bicuspid aortic valve can be passed down in families, meaning it's inherited. Parents, children and siblings of someone with a bicuspid aortic valve should have an echocardiogram to check for the condition.

**EPIDEMIOLOGY**

The epidemiology of aortic valve disease varies enormously between high-income and low-income countries. The majority of morbidity and mortality attributable to aortic valve disease worldwide is due to infectious disease. It may either be directly, as in infective endocarditis, or indirectly, as in acute rheumatic fever (ARF), which is most commonly seen in low-income countries. In high-income countries, the greatest burden of aortic valve disease referred to hospitals is due to degenerative calcific aortic valve disease.

The number of cases of aortic valvular disease will increase because of the strong association between valvular disease and age, combined with the rapid ageing of populations worldwide . Valvular disease afflicts the elderly in developed nations, it is insidious in onset, and frequently associated with other comorbidities. However, in low-income countries valvular disease is encountered in the young, not infrequently in children of school age or young females of child-bearing potential, and with a course that is much more rapid. The burden of rheumatic heart disease (RHD) falls disproportionately on low-income countries and in low-income groups in high-income countries. The scope and magnitude of cardiovascular disease are vastly different in different countries (Africa, Asia, Europe or North America).

The incidence of acute rheumatic fever has been difficult to establish globally. Estimates range from 10 cases per 100,000 to as high as 374 cases per 100,000 in Pacific and indigenous Australian and New Zealand communities . Indigenous Australians aged 5–14 years (the peak age group to develop ARF have an incidence of 194 per 100,000) . Similarly, in New Zealand, the overall population age-standardised incidence of 17.2 per 100,000 for ARF requiring hospitalisation masks an almost twentyfold increased rate for Maori (40 per 100,000) and a fortyfold increased rate in Pacific people (81 per 100,000) compared with New Zealanders of non-Maori/Pacific origin (2.1 per 100,000)

In Africa, ARF is still seen regularly in its most fulminant form, affecting children as young as six years old. It manifests with severe refractory heart failure, often requiring valve replacement in childhood. In the continent of Africa, the World Health Organization expert panel on rheumatic fever (RF) and RHD estimated that in 1994 about 12 million people worldwide had RF and RHD, most of them in developing countries . A more recent review of the current evidence for the global burden of RF and RHD in Africa, estimated that 15.6–19.6 million people have RHD (2.4 million children aged 5–14 years) causing 233,364 – 294,398 deaths from RHD each year (based on an annual mortality rate of 1.5%) [5,6]. Then, the highest prevalence of RHD is in sub-Saharan Africa with a prevalence of 5.7 per 1,000, compared with 1.8 per 1,000 in North Africa, and 0.3 per 1,000 in economically developed countries .

Recent studies from Africa, using echocardiographic screening, found subclinical RHD in children with a nearly tenfold higher prevalence than without echocardiographic study

While previous studies have estimated that the number of children aged 5-14 years with RHD in Asia is between 1.96 and 2.21 million, a recent meta-analysis, using echocardiographic diagnosed RHD, estimated that the prevalence of RHD in Southeast Asia was 28 per 1,000. In another study, the echocardiographic prevalence of RHD in Indian school children was 20 per 1,000. The fact that the observed prevalence of RHD in India is similar to sub-Saharan Africa suggests that economic betterment may not have translated to improvement in healthcare systems across India. A review of recent studies predominantly using echocardiography for the diagnosis of chronic RHD shows wide global variations in prevalence, between 46 per 100,000 in northern India and 2,400 per 100,000 in the Solomon Islands

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis for a bicuspid aortic valve includes the following:

* Coarctation of the aorta
* Interrupted aortic arch
* Pediatric aortic arch insufficiency
* Pediatric Ebstein anomaly
* Pediatric mitral valve prolapse
* Pediatric rheumatic heart disease
* Pediatric sub valvar aortic stenosis
* Turner syndrome
* Valvar pulmonary stenosis
* Williams syndrome

**COMPLICATION**

The bicuspid aortic valve can lead to significant valvular dysfunction which could be aortic stenosis or aortic regurgitation. The risk of infective endocarditis is high among populations with a bicuspid valve. The bicuspid aortic valve is also associated with:

* Coarctation of aorta
* Ascending aortic dilatation 40% to 60%
* Aortic aneurysm

REFERENCES

[Bicuspid aortic valve - Overview - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/bicuspid-aortic-valve/cdc-20385577)

[Epidemiology of aortic valve stenosis (AS) and of aortic valve incompetence (AI): is the prevalence of AS/AI similar in different parts of the world?](https://www.escardio.org/Journals/E-Journal-of-Cardiology-Practice/Volume-18/epidemiology-of-aortic-valve-stenosis-as-and-of-aortic-valve-incompetence-ai)

**Transposition of the great arteries**

Transposition of the great arteries (TGA) is a serious, rare heart problem in which the two main arteries leaving the heart are reversed. The condition is present at birth, which means it's a congenital heart defect.

### **Types**

There are two types of transposition of the great arteries:

* **Complete transposition of the great arteries**, also called dextro-transposition of the great arteries (D-TGA). This type reduces the amount of oxygen-rich blood to the body. Symptoms are usually noticed during pregnancy, immediately after birth or within a few weeks of birth. Without treatment, serious complications or death can occur.

d-TGA happens when the main pulmonary artery and the aorta are switched in position, or "transposed." The main pulmonary artery and the aorta are the two main arteries that carry blood out of the heart.

In babies with d-TGA, oxygen-poor blood from the body enters the right side of the heart. But, instead of going to the lungs, the blood is pumped back out to the body through the aorta. Oxygen-rich blood from the lungs entering the heart is pumped straight back to the lungs through the main pulmonary artery.

Often, babies with d-TGA have other heart defects, such as a ventricular septal defect (VSD) or an atrial septal defect (ASD). These defects allow blood to mix so that some oxygen-rich blood can be pumped to the rest of the body. The patent ductus arteriosus (labeled PDA in the image) also allows some oxygen-rich blood to be pumped to the rest of the body.

* **Congenitally corrected transposition**, also called levo-transposition of the great arteries (L-TGA). This is a less common type. Symptoms may not be noticed right away. Treatment depends on the specific heart problems.

Surgery to correct the positions of the arteries is the usual treatment. The surgery is usually done soon after birth.

**Causes**

Transposition of the great arteries occurs during pregnancy when the baby's heart is developing. The cause is most often unknown.

To understand transposition of the great arteries, it may be helpful to know how the heart typically pumps blood.

* Usually, the artery that carries blood from the heart to the lungs — called the pulmonary artery — connects to the heart's lower right chamber. That chamber is called the right ventricle.
* Oxygen-rich blood is then pumped from the lungs to the heart's upper left chamber, also called the left atrium.
* Blood then flows into the lower left chamber, known as the left ventricle.
* The body's main artery, called the aorta, typically connects to the left ventricle. It carries oxygen-rich blood out of the heart to the rest of the body.

### **Complete transposition of the great arteries (D-TGA)**

In a complete transposition of the great arteries (also called dextro-transposition of the great arteries), the two arteries leaving the heart have switched positions. The pulmonary artery connects to the left lower heart chamber. The aorta connects to the right lower heart chamber.

The switched arteries cause changes in blood flow. Oxygen-poor blood now flows through the right side of the heart. It goes back to the body without passing through the lungs. Oxygen-rich blood now flows through the left side of the heart. It goes directly back into the lungs without being pumped to the rest of the body.

### **Congenitally corrected transposition (L-TGA)**

In this less common type, also called levo-transposition of the great arteries (L-TGA), the two lower heart chambers are reversed.

* The lower left heart chamber, called the left ventricle, is on the heart's right side. It gets blood from the upper right heart chamber.
* The lower right heart chamber is on the heart's left side. It gets blood from the left upper heart chamber.

The blood usually still flows correctly through the heart and body. But the heart can have long-term trouble pumping blood. People with L-TGA may also have problems with the tricuspid heart valve.

**Risk factors**

Several things may increase a baby's risk of transposition of the great arteries, including:

* A history of German measles (rubella) or another infection by a virus during pregnancy.
* Drinking alcohol or taking certain medicines during pregnancy.
* Smoking during pregnancy.
* Poorly controlled diabetes during pregnancy.

**Symptoms**

Transposition of the great arteries (TGA) may be seen in a baby before birth during a routine pregnancy ultrasound.

But some people with the congenitally corrected type of TGA may not have symptoms for many years.

Symptoms of transposition of the great arteries after birth include:

## Blue or gray skin. Depending on the baby's skin color, these color changes may be harder or easier to see.

## Weak pulse.

## Lack of appetite.

## Poor weight gain.

## Skin color changes may not be noticed right away if a baby with TGA also has other heart problems. This is because the other heart problems may let some oxygen-rich blood move through the body. But as the baby becomes more active, less blood flows through the body. The blue or gray skin color then becomes more noticeable.

Symptoms for d-TGA occur at birth or very soon afterwards. How severe the symptoms are will depend on whether oxygen-rich blood can get out to the rest of the body. For example, if an infant with d-TGA has an atrial septal defect (ASD), the ASD forms a passageway for some oxygen-rich blood to reach the rest of the body. Therefore, this infant with both defects may not have as severe symptoms as infants whose hearts don't have any mixing of blood.

Infants with d-TGA can have a bluish looking skin color because their blood doesn't carry enough oxygen. This condition is called cyanosis. Infants with d-TGA or other conditions causing cyanosis can have symptoms such as

* Problems breathing
* Pounding heart
* Weak pulse
* Ashen or bluish skin color
* Poor feeding

**When to see a doctor**

Always seek emergency medical help if you notice that anyone develops a blue or gray skin color.

**Diagnosis**

Transposition of the great arteries is most often diagnosed after a baby is born. But sometimes the condition may be seen before birth during a routine pregnancy ultrasound. If so, an ultrasound of the unborn baby's heart may be done to confirm the diagnosis. This test is called a fetal echocardiogram.

After birth, a health care provider may think about a diagnosis of a TGA if the baby has blue or gray skin, a weak pulse, or trouble breathing. The care provider may hear a heart sound, called a murmur, while listening to the baby's heart.

**Tests**

Tests are needed to confirm a diagnosis of transposition of the great arteries. They may include:

## Echocardiogram. This test uses sound waves to create moving pictures of the beating heart. It shows how blood flows through the heart, heart valves and blood vessels. It can show the positions of the two main arteries leaving the heart. An echocardiogram also can show if there are other heart problems present at birth, such as a hole in the heart.

## Chest X-ray. A chest X-ray shows the condition of the heart and lungs. It can't diagnose TGA by itself, but it does help the health care provider see the heart's size.

## Electrocardiogram (ECG or EKG). This simple, painless test records the electrical activity of the heart. Sticky patches called electrodes are placed on the chest and sometimes the arms and legs. Wires connect the electrodes to a computer, which displays the test results. An ECG can show if the heart is beating too fast, too slow or not at all.

## **Treatment**

## All infants with complete transposition of the great arteries (D-TGA) need surgery to correct the heart problem. Treatment for congenitally corrected transposition (L-TGA) depends on when the condition is diagnosed and what other heart conditions exist.

## **Medications**

## Before surgery is done to fix the switched arteries, a medicine called alprostadil (Caverject, Edex, others) may be given to the baby. This medicine increases blood flow. It helps oxygen-poor and oxygen-rich blood better mix together.

## Surgery or other procedures

## Surgery for transposition of the great arteries (TGA) is usually done within the first days to weeks after birth. Options depend on the type of TGA. Not all people with congenitally corrected transposition need surgery.

## Surgeries and other treatments used to treat transposition of the great arteries may include:

## Atrial septostomy. This treatment may be done urgently as a temporary fix before surgery. It uses thin tubes and small cuts to widen a natural connection between the heart's upper chambers. It helps mix oxygen-rich and oxygen-poor blood, improving oxygen levels in the baby's body.

## Arterial switch operation. This is the most common surgery used to correct transposition of the great arteries. During this surgery, the two main arteries leaving the heart are moved to their correct positions. Other heart problems present at birth may be repaired during this surgery.

## Atrial switch operation. The surgeon splits blood flow between the heart's two upper chambers. After this surgery, the right lower heart chamber must pump blood to the body, instead of just to the lungs.

## Rastelli procedure. This surgery may be done if a baby with TGA also has a hole in the heart called a ventricular septal defect. The surgeon patches the hole and redirects blood flow from the left lower heart chamber to the aorta. This lets oxygen-rich blood go to the body. An artificial valve connects the right lower heart chamber to the lung artery.

## Double switch procedure. This complex surgery is used to treat congenitally corrected transposition. It redirects blood flow coming into the heart. It switches the great artery connections so the left lower heart chamber can pump oxygen-rich blood to the aorta.

## Babies born with TGA often have other heart problems. Other surgeries may be needed to fix those heart problems. Surgery also may be needed to treat complications of TGA. If TGA causes changes in the heartbeat, a device called a pacemaker may be recommended.

## After surgery to fix TGA, lifelong care is needed with a provider trained in heart problems present at birth. This type of health care provider is called a congenital cardiologist.

## **Complications**

Complications depend on the type of transposition of the great arteries (TGA).

Possible complications of complete transposition of the great arteries (D-TGA) may include:

* **Not enough oxygen to body tissues.** Unless there's some mixing of oxygen-rich blood and oxygen-poor blood within the body, this complication causes death.
* **Heart failure.** Heart failure is a condition in which the heart can't pump enough blood to meet the body's needs. It may develop over time because the right lower heart chamber is pumping under higher pressure than usual. The strain may make the muscle of the right lower chamber stiff or weak.

Possible complications of congenitally corrected transposition (L-TGA) may include:

* **Reduced heart pumping.** In L-TGA, the right lower heart chamber pumps blood to the body. This work is different from what that chamber was designed to do. This can cause changes in how well the heart pumps blood.
* **Complete heart block.** The changes in the structure of the heart due to L-TGA can change the electrical signals that tell the heart to beat. A complete heart block occurs if all signals are blocked.
* **Heart valve disease.** In L-TGA, the valve between the upper and lower heart chambers — the tricuspid valve — may not close completely. Blood might move backward through the valve. This condition is called tricuspid valve regurgitation. It can eventually reduce the heart's ability to pump blood.

### **Pregnancy and TGA**

If you have transposition of the great arteries and want to become pregnant, talk with a health care provider first. It may be possible to have a healthy pregnancy, but special care may be needed.

Complications of TGA, such as changes in heart signaling or serious heart muscle problems, may make pregnancy risky. Pregnancy isn't recommended for people who have severe complications of TGA, even if they had surgery to fix the TGA.

**Prevention**

If you have a family history of heart problems present at birth, consider talking with a genetic counselor and a health care provider experienced in congenital heart defects before getting pregnant.

It's important to take steps to have a healthy pregnancy. Before becoming pregnant, get recommended immunizations and start taking a multivitamin with 400 micrograms of folic acid.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnoses for TGA include the following conditions:

* Double-outlet right ventricle
* Tricuspid atresia
* Pulmonary atresia
* Tetralogy of Fallot
* Total anomalous pulmonary venous return
* Truncus arteriosus

A thorough clinical investigation and appropriate imaging studies are essential to differentiate TGA from these conditions and guide management.

**PROGNOSIS**

Surgical repair for d-TGA should be performed within the first week of life. The two most commonly used procedures are the standard ASO and the Rastelli procedure, which is recommended for patients with d-TGA, a large VSD, and pulmonary stenosis. Studies report a survival rate exceeding 95% at 15 to 25 years post-discharge. Other corrective procedures, such as the Mustard, Senning, Nakaidoh, and REV, are available but less commonly performed.

Postoperative complications can result from the underlying pathophysiology, surgical intervention, or residual defects, leading to adverse effects such as right ventricular dysfunction, tricuspid valve regurgitation, supraventricular arrhythmias, interatrial and interventricular septal dysfunction, and, less commonly, pulmonary hypertension. Despite these potential complications, most treated patients reach adulthood, with a 20-year survival rate nearing 90%. The primary cause of death is sudden cardiac death, followed by anatomical right ventricular dysfunction.

Significant concerns exist regarding the ability of the anatomical right ventricle to sustain systemic circulation in patients with TGA who have undergone the Mustard procedure. In this interatrial procedure, the anatomical right ventricle becomes the systemic ventricle, responsible for pumping blood into the main artery and managing pressures 3 to 4 times higher than those in the pulmonary circuit, leading to a considerable pressure load. Over time, the anatomical right ventricle often struggles to maintain systemic circulation, leading to a gradual decline in the clinical condition of patients following Mustard repair. These late complications are associated with poor outcomes, emphasizing the importance of timely intervention.

Additionally, conduction and electrical stimulation disorders are common complications in adults who have undergone interatrial surgery for d-TGA. These issues may arise from congenital anomalies of the sinus node and conductive fibers, damage to these structures during surgery, or injury to the coronary arteries supplying these areas. Over time, the inevitable degeneration of tissues also contributes to these complications. Notably, only 40% to 50% of patients who underwent the Senning procedure maintain sinus rhythm after 20 years of follow-up.

In ccTGA, the improved long-term survival associated with anatomic repair must be carefully weighed against the short-term safety of a physiological repair strategy, which focuses solely on correcting associated abnormalities.However, surgical mortality in the physiological repair population is not insignificant. For instance, a series from the Mayo Clinic reported a 3% surgical mortality rate for operations performed after 1986, with an overall mortality rate of 16%. Similarly, a Dutch series by Bogers et al documented a 6.7% mortality rate. An alternative approach for managing cc-TGA is staged single-ventricle palliation with Fontan completion. While short-term outcomes regarding survival and symptoms are comparable, the long-term complications are significant, with many patients experiencing serious morbidities.

Reinterventions after anatomic correction for cc-TGA are common. In particular, conduit replacements are especially frequent in the atrial switch-Rastelli cohort, with most patients likely requiring at least one exchange to a larger conduit suitable for transcatheter replacement. After the initial reoperation, procedures such as multiple transcatheter replacements are often performed, reducing the need for further surgeries. Complications related to baffles, such as residual leaks or stenosis, are well-documented after Mustard or Senning operations and are a common reason for reoperations following anatomic repair for ccTGA. However, in the current era, most baffle-related issues can be effectively managed with transcatheter techniques, minimizing the need for additional surgeries. Residual LVOTO is also a known complication following both types of anatomic repair.

**COMPLICATION**

Untreated TGA can result in CHF, life-threatening arrhythmias, and death. Complications that may arise from corrective procedures for TGA include:

* Arrhythmias
* Obstruction or leakage of the baffle (following a Rastelli procedure)
* Pulmonary artery stenosis
* Coronary artery stenosis
* Aortic root dilation
* Aortic regurgitation

Timely intervention and regular follow-up care can help minimize complications and improve outcomes for patients with TGA.

**EPIDEMIOLOGY**

D-TGA is a CHD occurring in about 1 in 4000 live births. Although some cases of d-TGA have been linked to rare variants in specific genes, the genetic basis for most cases remains unclear. The pattern of familial recurrence, along with the sporadic nature of the majority of d-TGA cases, suggests polygenic inheritance. TGA accounts for 3% of all CHDs and 20% of cyanotic heart diseases.

CcTGA is an uncommon and complex CHD, occurring in about 0.05% of all CHD cases, with an estimated incidence of approximately 1 in every 33,000 live births. An international study examining the largest cohort of ccTGA cases identified non sporadic occurrences, including familial clusters. The study also found associations between ccTGA, d-TGA, laterality defects, and, in some instances, primary ciliary dyskinesia. These findings suggest a potential common pathogenetic pathway involving laterality genes in the development of ccTGA.

CcTGA is more prevalent in male infants. Due to the physiologically corrected circulation, isolated ccTGA typically remains asymptomatic during childhood and may go unnoticed until the sixth decade of life. At this stage, ccTGA can present with right ventricular dysfunction and heart block. Approximately 90% of individuals with ccTGA have additional cardiac abnormalities that affect clinical symptoms and prognosis. Commonly associated conditions include VSD in 80% of cases, pulmonary stenosis in 40% to 50%, left ventricular outflow tract obstruction (LVOTO) in 30%, and Ebstein anomaly in 30%. Other potential anomalies include situs inversus, dextrocardia, complete heart block, and reentrant tachycardias. Symptoms may manifest as bradycardia, fatigue, and reduced exercise tolerance

REFERENCES

[Transposition of the great arteries - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/transposition-of-the-great-arteries/symptoms-causes/syc-20350589)

[Transposition of the great arteries - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/transposition-of-the-great-arteries/diagnosis-treatment/drc-20350595)

<https://www.cdc.gov/heart-defects/about/d-tga.html>

[Transposition of the Great Arteries - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK538434/#article-30506.s4)

**Hypoplastic left heart syndrome (HLHS)**

Hypoplastic left heart syndrome (HLHS) is a rare heart condition that a child is born with. That means it's a congenital heart defect. In this condition, the left side of the heart doesn't develop fully and is too small. So it can't pump blood well. Instead, the right side of the heart must pump blood to the lungs and to the rest of the body.

Treatment for hypoplastic left heart syndrome may include medicines, heart surgery or a heart transplant. Advances in care have improved the outlook for babies born with HLHS.

**CAUSES**

Hypoplastic left heart syndrome (HLHS) happens in the womb when a baby's heart develops. The cause isn't known. Gene changes may play a role.

In hypoplastic left heart syndrome, the left side of the heart hasn't grown enough so it does not develop fully. It can't properly send blood to the body. In HLHS, the following areas of the heart are too small:

## The lower left heart chamber, called the left ventricle.

## The body's main artery, called the aorta.

## The heart valves on the left side of the heart, called the aortic and mitral valves.

## After birth, the right side of a baby's heart usually pumps blood both to the lungs and to the rest of the body. The blood passes through an opening called the ductus arteriosus. This opening, also called a vessel, connects the pulmonary artery directly to the aorta. The oxygen-rich blood goes back to the right side of the heart through a natural opening between the upper chambers of the heart. The opening is called the foramen ovale.

## The ductus arteriosus usually closes after the first day or two of life. When that happens, the right side of the heart has no way to pump blood to the body. The left side of the heart takes over this job.

## But in babies with hypoplastic left heart syndrome, the left side can't pump blood well. So they need medicine to keep these connections open and keep blood flowing to the body until they have heart surgery.

## **Risk factors**

## People who have a child with hypoplastic left heart syndrome (HLHS) have a higher risk of having another baby with this or a similar condition.

## There are no other clear risk factors for hypoplastic left heart syndrome.

## **Symptoms**

Babies born with hypoplastic left heart syndrome (HLHS) usually are very sick soon after birth. Symptoms of HLHS include:

* Blue or gray skin, lips or fingernails. Depending on skin color, these changes may be harder or easier to see.
* Rapid, difficult breathing.
* Poor feeding.
* Cold hands and feet.
* Weak pulse.
* Being more drowsy or less active than is typical for most babies.

Without treatment, a baby with this condition may go into shock. Symptoms of shock include:

* Cool, clammy skin that can be pale or lips that can be blue or gray.
* A weak and rapid pulse.
* Breathing that may be slow and shallow or very rapid.
* Dull eyes that seem to stare

## **Diagnosis**

To diagnose hypoplastic left heart syndrome (HLHS), a healthcare professional examines the baby and listens to the baby's heart. The healthcare professional may hear a sound called a heart murmur. Rushing blood flow causes this sound.

### **Tests**

Tests used to find hypoplastic left heart syndrome (HLHS) in the baby before or after birth may include:

* **Pregnancy ultrasound.** A routine ultrasound exam during the second trimester of pregnancy usually can tell if the baby has HLHS.
* **Echocardiogram.** This test uses sound waves to make pictures of the heart. It shows how blood flows through the heart. It can be used after a baby is born to diagnose hypoplastic left heart syndrome. If a baby has HLHS, the test might find that the lower left heart chamber and heart valves are small. The body's main artery, called the aorta, also may be small.

**Treatment**

A baby born with hypoplastic left heart syndrome (HLHS) needs urgent treatment. Treatment can include many surgeries or a heart transplant. Medicines and other therapies are used to manage symptoms before heart surgery.

Talk with your child's healthcare professional about treatment options for your child.

If hypoplastic left heart syndrome is found before birth, healthcare professionals usually recommend giving birth at a hospital with a cardiac surgery center.

### **Medications**

The medicine alprostadil (Prostin VR Pediatric) is used to keep the ductus arteriosus open. It typically closes in all babies soon after birth. But in babies with hypoplastic left heart syndrome, the ductus needs to stay open so that blood can go to the rest of the body.

### **Therapies**

While waiting for surgery or a heart transplant, a baby with hypoplastic left heart syndrome may be given medicine and have these treatments:

* **Breathing help.** Babies who have trouble breathing may need help from a breathing machine called a ventilator.
* **Fluids through a vein.** A baby might receive fluids through a tube inserted into a vein. These are called intravenous (IV) fluids.
* **Feeding tube.** Babies who have trouble feeding or who tire while feeding can be fed through a feeding tube.

### **Surgery or other procedures**

Most children with hypoplastic left heart syndrome need several surgeries.

* **Atrial septostomy.** This treatment uses tubes called catheters and a balloon to make or widen an opening between the heart's upper chambers. It lets more blood flow between the upper two chambers of the heart. This treatment is done if the foramen ovale closes or is too small. Babies born with a hole in the heart, called an atrial septal defect, might not need atrial septostomy.

Other surgeries can make separate pathways to get the correct blood flow to the body and lungs. The surgeries are done in three stages.

* **Norwood procedure.** This surgery is often done within the first two weeks of life. There are various ways to do this treatment.  
  Surgeons rebuild the aorta and connect it to the heart's lower right chamber. Then they add a tube called a shunt to provide the lungs with blood. The surgeons may use one of two types of tubes. One type of tube connects the body's main artery to the arteries leading to the lungs. Those are the pulmonary arteries. The other type of tube goes from the right lower heart chamber to the pulmonary arteries. This treatment lets the right lower heart chamber pump blood to both the lungs and the body.  
  Sometimes, a mixed, also called hybrid, procedure is done. Heart surgeons place a stent in the ductus arteriosus to maintain the opening between the pulmonary artery and the aorta. Then they place bands around the pulmonary arteries to reduce blood flow to the lungs. They also make an opening between the upper chambers of the heart.  
  After the Norwood procedure, a baby's skin often is still slightly still slightly blue or gray. This is because oxygen-rich and oxygen-poor blood continue to mix within the heart. The blue or gray color may be easier or harder to see in some babies. Once a baby has this treatment, the odds of survival can go up.
* **Bidirectional Glenn procedure.** This tends to be the second surgery. It's usually done when a child is between 4 and 6 months of age. It involves removing the first shunt and connecting the large vein that drains blood from the head and arms to the pulmonary artery. Now the lungs receive blood from the vein instead of the shunt. The large vein is called the superior vena cava.  
  This surgery lessens the work of the right lower heart chamber by letting it pump blood mainly to the aorta. It also lets most of the oxygen-poor blood returning from the body flow directly into the lungs. After this treatment, all the blood returning from the upper body flows to the lungs. So blood with more oxygen is pumped to the aorta to supply organs and tissues throughout the body.
* **Fontan procedure.** This surgery usually is done when a child is between 3 and 4 years of age. The surgeon creates a path for the blood from the lower legs to flow directly into the pulmonary arteries. The vessel involved is called the inferior vena cava. The pulmonary arteries then send the blood into the lungs.  
  The Fontan procedure lets the rest of the oxygen-poor blood returning from the body flow to the lungs. After this surgery, there's little mixing of oxygen-rich and oxygen-poor blood in the heart. So the skin should no longer look blue or gray.
* **Heart transplant.** Many babies with hypoplastic left heart syndrome need a heart transplant. (8) Children who have heart transplants need to take medicines for life so that their bodies don't reject the donor heart.

### **Follow-up care**

After surgery or a transplant, a baby needs lifelong care with a heart doctor trained in congenital heart disease to watch for complications. Your child may need further treatment or other medicines for these complications.

Some children may need to take antibiotics before certain dental or other procedures to help prevent infections. Ask your child's healthcare professional if your child needs preventive antibiotics. Some children also may need to limit physical activity.

### **Follow-up care for adults**

Adults who were born with hypoplastic left heart syndrome (HLHS) need to see a heart doctor trained in congenital heart disease in adults. Recent advances in surgical care have helped children with HLHS grow into adulthood. So it's not yet clear what challenges an adult with the heart condition might have. Adults need regular, lifelong follow-up care to watch for changes in the condition.

People thinking about becoming pregnant should talk with their healthcare professionals about pregnancy risks and birth control options. Having hypoplastic left heart syndrome raises the risk of:

* Heart and blood vessel problems during pregnancy.
* Miscarriage.
* A baby being born with a congenital heart defect.

**Complications**

### With proper treatment, many babies with hypoplastic left heart syndrome (HLHS) survive. But they do need many surgeries and can have less energy and other challenges. Complications of HLHS may include:

### Getting tired more easily during sports or other exercises.

### Irregular heartbeats, called arrhythmias.

### Fluid buildup, called edema, in the lungs, stomach area, legs and feet.

### Not growing well.

### Developmental conditions related to the brain and nervous system.

### Need for more heart surgery or a heart transplant.

### **Prevention**

### There's no way to prevent hypoplastic left heart syndrome. If you were born with a heart condition, talk with a heart doctor and genetic counselor before getting pregnant.

### **When to see a doctor**

Get emergency medical help if your baby has:

* Changes in skin or nail color.
* Trouble breathing or fast breathing.
* Weak pulse or rapid pulse.
* Cool clammy skin.

**DIFFERENTIAL DIAGNOSIS**

Several diseases or conditions can mimic the presentation of HLHS, particularly in neonates with cyanosis, signs of poor perfusion, or congestive heart failure. Accurate differentiation is critical, as management strategies for these conditions differ significantly from HLHS. Below is a discussion of diseases or conditions that can be mistaken for HLHS:

* Critical aortic stenosis
  + Severe aortic stenosis can cause LV hypertrophy and poor systemic perfusion, resembling HLHS. However, the LV is typically present in critical aortic stenosis and may be dilated rather than hypoplastic. Echocardiography can differentiate this condition by visualizing the LV size and function and assessing the degree of aortic valve obstruction.
* Coarctation of the aorta
  + Coarctation of the aorta, especially with a large ventricular septal defect, can lead to cyanosis and shock as the ductus arteriosus begins to close. Because of systemic hypoperfusion, this condition may be confused with HLHS, but echocardiography will show a normally sized or hypertrophied LV and an obstruction localized to the aortic isthmus.
* Shone complex
  + Shone complex is a rare congenital heart condition characterized by multiple levels of left-sided obstructive lesions, including a supravalvular mitral membrane, a parachute mitral valve with all chordae attached to a single papillary muscle, subaortic stenosis caused by a fibromuscular ridge below the aortic valve, and coarctation of the aorta impairing systemic blood flow. This condition may be mistaken for HLHS because both conditions involve reduced left-sided cardiac output and systemic hypoperfusion. Symptoms such as cyanosis, poor perfusion, and heart failure, as the ductus arteriosus closes, are common to both, but echocardiography can differentiate them by identifying the discrete levels of obstruction in the Shone complex versus the global underdevelopment of the left heart in HLHS.
* Interrupted aortic arch
  + This condition can present with systemic hypoperfusion and shock, similar to HLHS. This condition involves complete discontinuity of the aortic arch, but echocardiography or advanced imaging will reveal a normal or mildly hypoplastic LV and the specific site of aortic interruption.
* Total anomalous pulmonary venous return (TAPVR)
  + In TAPVR with obstruction, neonates may present with cyanosis and signs of low cardiac output that mimic HLHS. However, TAPVR is characterized by abnormal pulmonary venous drainage into the RA or systemic veins, which can be identified on echocardiography. The left-sided structures are typically normal in size and morphology.
* Pulmonary atresia with intact ventricular septum (PA/IVS)
  + PA/IVS may resemble HLHS due to ductal-dependent systemic perfusion and cyanosis. However, in PA/IVS, the LV is usually normal in size, and the obstruction is at the pulmonary valve rather than the aortic or mitral valves.
* Ebstein anomaly of the tricuspid valve
  + Severe forms of Ebstein anomaly can cause cyanosis and right-sided heart failure, potentially mimicking HLHS. However, echocardiography will show significant apical displacement of the tricuspid valve with atrialization of the RV, distinguishing this condition from HLHS.

**PROGNOSIS**

One-third of neonates with HLHS die before undergoing any palliative surgical intervention. The pre-Fontan mortality rate is approximately 70%, whereas post-Fontan mortality decreases to around 5%. Neonates with a single ventricle face a heightened risk of cardiac arrest (12.7%) and a mortality rate of 62.3%. Some centers report survival rates as high as 90% following stage 1 Norwood palliation, but only two-thirds of children with HLHS survive to age 5, with an annual mortality rate of approximately 1% among Fontan patients. The 2019 Society of Thoracic Surgeons Congenital Heart Surgery Database reported stage 1 palliation mortality at 15%, hemi-Fontan/Glenn at 1.8%, and Fontan at 1.0%.

The Aristotle score is a system used to predict the survival chances of patients with congenital heart disease. This system calculates the risk of early mortality and morbidity, such as the length of stay in the intensive care unit, while also considering the anticipated difficulty of the surgical technique. This scoring system evaluates the complexity of the surgical procedure and assigns additional points based on the patient's current health condition. Points may be added for various factors, including anatomical variations, respiratory failure, shock, prematurity, and low weight. A score of 20 or higher is associated with a high mortality rate during surgery, and such scores are often observed in patients undergoing the Norwood procedure. The Aristotle score is calculated before surgery to help counsel parents and provide them with realistic expectations about the procedure.

Thirty-five years after the first Norwood operation, adults with hypoplastic left heart syndrome (HLHS) represent a small but growing population; these individuals frequently exhibit reduced aerobic capacity, which tends to decline progressively with age. Morbidity and mortality in this population are frequently associated with complications such as heart failure, PLE, significant thrombotic events, malignant arrhythmias, and, in some cases, the need for heart transplantation.

**Neurodevelopmental Issues**

Children with complex congenital cardiac diseases, particularly those with HLHS, face a higher risk of developmental delays, disabilities, and behavioral issues. Fetuses with HLHS exhibit decreased brain volume and metabolism, partially due to diminished cerebral oxygen delivery and utilization. Older infants and toddlers with HLHS often exhibit developmental delays, with Psychomotor Development Index scores consistently lower than Mental Development Index scores. A specific area of concern is visual-motor integration, which shows notable delays.

However, children with HLHS who have undergone staged Fontan palliation can achieve normal developmental outcomes. Nonetheless, they remain at significant risk for learning disorders, lower academic performance, and behavioral challenges. Various factors likely influence neurodevelopment in these children, including associated neurological malformations, genetic conditions, anatomical features, surgical techniques, and intraoperative perfusion methods.

**EPIDEMIOLOGY**

HLHS is a rare congenital heart defect, accounting for approximately 2% to 3% of all congenital heart defects and about 6% to 9% of critical congenital heart disease requiring surgery or catheter-based intervention in the neonatal period. The incidence of HLHS is estimated to be 0.16 to 0.36 per 1000 live births and accounts for 1.4% to 3.8% of all congenital heart defects. Males are affected more frequently than females, with a male-to-female ratio of approximately 1.5:1. HLHS is responsible for 23% of cardiac deaths during the first week of life, highlighting its significant contribution to early neonatal mortality.

Several study results have suggested a multifactorial etiology involving genetic and environmental factors. Although most cases of HLHS are sporadic, up to 20% may occur in association with chromosomal abnormalities or syndromes, such as Turner syndrome, trisomy 13, or trisomy 18. Familial clustering has been observed, and mutations in genes like *NOTCH1* and *HAND1* have been implicated, suggesting a genetic predisposition in some cases.

HLHS is often diagnosed prenatally through routine fetal ultrasonography, with advances in imaging techniques enabling earlier and more accurate detection. Early diagnosis has improved perinatal management and outcomes. Despite its rarity, HLHS remains a leading cause of neonatal mortality due to congenital heart disease if left untreated, underscoring the importance of early intervention and specialized care.

REFERENCES

[Hypoplastic left heart syndrome - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/hypoplastic-left-heart-syndrome/symptoms-causes/syc-20350599)

[Hypoplastic left heart syndrome - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/hypoplastic-left-heart-syndrome/diagnosis-treatment/drc-20350605)

[Hypoplastic Left Heart Syndrome - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK554576/#article-23285.s4)

**Tricuspid atresia**

Tricuspid atresia occurs when the tricuspid valve in the heart does not form at all. The tricuspid valve controls blood flow from the right atrium to the right ventricle. The right atrium is the upper right chamber of the heart. The right ventricle is the lower right chamber of the heart.

In babies with tricuspid atresia, blood can’t flow correctly through the heart and to the rest of the body. Blood is unable to get from the right atrium through the right ventricle and out to the lungs. For this reason, the right ventricle can be underdeveloped. The main pulmonary artery may also be small with very little blood going through it to the lungs.

A baby with tricuspid atresia may need surgery or other procedures soon after birth. Therefore, tricuspid atresia is considered a critical congenital heart defect (critical CHD).

People with tricuspid atresia can't get enough oxygen through the body. So they tire easily and they are often short of breath. Their skin and lips may look blue or gray.

Tricuspid atresia is treated with multiple surgeries. Most babies with tricuspid atresia who have surgery live well into adulthood, though follow-up surgeries are often needed.

Other names for tricuspid atresia are:

* Tricuspid valve atresia
* TV atresia

## 

## **Causes**

Most congenital heart defects, including tricuspid atresia, result from changes that occur early as the baby's heart is developing before birth. The exact cause is usually unknown.

### **How the heart works**

To understand more about tricuspid atresia, it may be helpful to know how the heart typically works.

The heart is divided into four chambers, two on the right and two on the left.

* The right upper chamber is called the right atrium.
* The right lower chamber is called the right ventricle.
* The left upper chamber is called the left atrium.
* The left lower chamber is called the left ventricle. The left ventricle is the heart's main pumping chamber.

To pump blood throughout the body, the heart uses its left and right sides for different tasks.

* The right side of the heart moves blood to the lungs through the lung arteries. These are the pulmonary arteries.
* In the lungs, blood picks up oxygen and then returns to the heart's left side through the pulmonary veins.
* The left side of the heart pumps the blood through the body's main artery, called the aorta. The blood then flows out to the rest of the body.

Valves control the flow of blood into and out of the heart. The heart valves open to allow blood to move to the next chamber or to one of the arteries. The heart valves close to keep blood from flowing backward.

### **What happens in tricuspid atresia**

In tricuspid atresia, the tricuspid valve is missing. A sheet of tissue blocks the flow of blood between the right heart chambers. There's no way for blood to move from the right upper chamber to the right lower chamber. The right side of the heart is no longer able to pump blood to the lungs.

Instead, blood flows from the heart's right upper chamber directly to the left upper chamber through a hole in the wall between them. The hole is either a congenital heart defect called an atrial septal defect or a natural opening called the foramen ovale. When the foramen ovale doesn't close after birth, it's called a patent foramen ovale.

How blood flows after that depends on whether there are other heart structure problems. In some babies with tricuspid atresia, blood moves from the left lower heart chamber into the body's main artery, the aorta. Then it goes to the lungs through a temporary connection called the ductus arteriosus. This connection typically closes after birth.

Many babies born with tricuspid atresia also have a hole between the lower heart chambers. This hole is called a ventricular septal defect (VSD). If this occurs, some blood can flow through the hole directly to the main lung artery. The amount of blood going to the lungs depends on the size of the VSD and whether the pulmonary valve is narrowed. If the VSD is large, too much blood can move to the lungs. This can cause heart failure.

**Risk factors**

It's not entirely clear why congenital heart defects such as tricuspid atresia occur. But some risk factors have been identified. Many babies born with a genetic disorder called Down syndrome have tricuspid atresia.

Other things that might increase your baby's risk of tricuspid atresia include:

* Having German measles (rubella) or another viral illness during early pregnancy
* Family history of congenital heart disease
* Drinking alcohol during pregnancy
* Smoking before or during pregnancy
* Poorly controlled diabetes during pregnancy
* Use of certain medications during pregnancy, including some used to treat acne, bipolar disorder and seizures

## **Symptoms**

Tricuspid atresia symptoms are usually seen soon after birth. Symptoms of tricuspid atresia may include:

* Blue or gray skin and lips due to low blood oxygen levels
* Difficulty breathing
* Tiring easily, especially during feedings
* Slow growth and poor weight gain

Some people with tricuspid atresia also develop symptoms of heart failure. Heart failure symptoms include:

* Fatigue and weakness
* Shortness of breath
* Swelling in the legs, ankles and feet
* Swelling of the belly area, a condition called ascites
* Sudden weight gain from a buildup of fluid

## 

## **Diagnosis**

## Tricuspid atresia may be diagnosed during a routine pregnancy ultrasound before a baby is born. It's important to get proper prenatal care during pregnancy.

## After birth, a health care provider immediately examines the baby and listens to the baby's heart and lungs. The care provider might suspect a heart problem such as tricuspid atresia if a baby has blue or gray skin, trouble breathing, or an irregular heart sound called a heart murmur. Changes in blood flow to and from the heart can cause a heart murmur.

### Tests

## Tests to diagnose tricuspid atresia may include:

## Echocardiogram. Sound waves create moving images of blood flow through the heart and heart valves. In a baby with tricuspid atresia, the echocardiogram shows a missing tricuspid valve and irregular blood flow. The test may reveal other heart problems as well.

## Electrocardiogram. Also called an ECG or EKG, this quick and painless test records the electrical activity of the heart. It can show how fast or how slow the heart is beating. An ECG can detect irregular heart rhythms.

## Pulse oximetry. A small sensor attached to a hand or foot measures the amount of oxygen in the blood. Pulse oximetry is simple and painless.

## Chest X-ray. A chest X-ray shows the condition of the heart and lungs. It can help determine the size of the heart and its chambers. A chest X-ray can show fluid buildup in the lungs.

## Cardiac catheterization. A thin, flexible tube called a catheter is inserted into a blood vessel, usually in the groin area, and guided into the heart. Dye flows through the catheter into the heart chambers. The dye helps the chambers be seen on X-ray images. The catheter can also be used to measure pressures in the heart chambers. Cardiac catheterization is rarely used to diagnose tricuspid atresia, but it might be done to examine the heart before tricuspid atresia surgery.

## 

## **Treatment**

## There's no way to replace a tricuspid valve in tricuspid atresia. If your child has tricuspid atresia, several surgeries are often done to improve blood flow through the heart and to the lungs. Medications are used to manage symptoms.

## If your baby has tricuspid atresia, consider getting care at a medical center with surgeons and other health care providers who have experience with complex congenital heart disease.

### Medications

## Medications for tricuspid atresia may be given to:

## Strengthen the heart muscle

## Lower blood pressure

## Remove excess fluid from the body

## Supplemental oxygen might be given to help the baby breathe better.

## Before heart surgery, a baby with tricuspid atresia may be given the hormone prostaglandin to help widen and keep open the ductus arteriosus.

### Surgeries or other procedures

## A baby with tricuspid atresia often needs several heart surgeries or procedures. Some of them are temporary fixes to quickly improve blood flow before a more permanent procedure can be done.

## Surgeries or procedures for tricuspid atresia include open-heart surgery and minimally invasive heart surgery. The type of heart surgery depends on the specific congenital heart defect.

## Shunting. This procedure creates a new pathway (shunt) for blood to flow. In tricuspid atresia, the shunt redirects blood from a main blood vessel leading out of the heart to the lungs. Shunting increases the amount of blood flow to the lungs. It helps improves oxygen levels. Surgeons generally place a shunt during the first two weeks of life. However, babies usually outgrow the shunt. They may need another surgery to replace it.

## Glenn procedure. In the Glenn procedure, the surgeon removes the first shunt. Then one of the large veins that typically returns blood to the heart is connected directly to the lung artery instead. The Glenn procedure reduces the strain on the heart's lower left chamber, decreasing the risk of damage to it. The procedure can be done when the pressures in the baby's lung have lowered, which happens as the baby gets older. The Glenn procedure sets the stage for a more permanent corrective surgery called the Fontan procedure.

## Fontan procedure. This type of heart surgery is typically done when a child is 2 to 5 years old. It creates a pathway so that most, if not all, of the blood that would have gone to the right heart can instead flow directly into the pulmonary artery. The short- and intermediate-term outlook for babies who have a Fontan procedure is generally promising. But regular checkups are necessary to monitor for complications, including heart failure.

## Pulmonary artery band placement. This procedure may be done if a baby with tricuspid atresia has a ventricular septal defect. The surgeon places a band around the main lung artery to reduce the amount of blood moving from the heart into the lungs.

## Atrial septostomy. Rarely, a balloon is used to create or enlarge the opening between the heart's upper chambers. This allows more blood to flow from the right upper chamber to the left upper chamber.

## After treatment, babies with tricuspid atresia need regular health checkups, ideally with a children's doctor trained in congenital heart conditions. This care provider is called a pediatric congenital cardiologist. Many children with congenital heart defects, such as tricuspid atresia, grow up to lead full lives.

## Adults treated for tricuspid atresia also need lifelong checkups, preferably with a doctor trained in adult congenital heart conditions. This care provider is called an adult congenital cardiologist.

## 

## **Self care**

If your child has tricuspid atresia, lifestyle changes may be recommended to keep the heart healthy and prevent complications.

Try these tips to help a baby or child with tricuspid atresia:

* **Adjust feedings.** A baby with tricuspid atresia might not be getting enough calories because of tiring during feeding and other factors. Try giving the baby frequent, small feedings.  
  Breast milk is an excellent source of nutrition. But a special high-calorie formula may be needed if your baby isn't getting enough nutrition because of tiring during feeding. Some babies might need to be fed through a feeding tube.
* **Ask about preventive antibiotics.** Sometimes, a congenital heart defect can increase the risk of infection in the lining of the heart or heart valves. This condition is called infective endocarditis. Antibiotics may be recommended before dental and other procedures to prevent this infection. Ask your child's heart doctor if preventive antibiotics are necessary for your child.  
  Practicing good oral hygiene — brushing and flossing teeth, getting regular dental checkups — also is important for good overall health.
* **Stay active.** Physical activity is important for heart health. Encourage as much play and activity as you or your child can tolerate or as your provider recommends. Allow lots of time for rest.
* **Discuss sports restrictions.** Some kids and adults with congenital heart defects may need to limit certain types of exercise or sports activities. A care provider can tell you if there are sports or activities that you or your child should limit or avoid.
* **Get recommended vaccines.** Standard immunizations are encouraged for children with congenital heart defects. So are vaccines for flu, COVID-19, pneumonia and respiratory syncytial virus infections.
* **Keep follow-up appointments with the care provider.** Your child will need at least yearly appointments with a pediatric congenital cardiologist.

### **Pregnancy and tricuspid atresia**

If you have tricuspid atresia and are pregnant or hoping to be, consider talking to an adult congenital heart disease specialist and a maternal-fetal medicine specialist. During pregnancy, it's important to receive care from a provider who specializes in pregnancies in those with congenital heart disease.

Pregnancy is considered high risk for those who have had a Fontan procedure. If you have a history of heart failure, you may be discouraged from becoming pregnant.

## **Complications**

Tricuspid atresia restricts blood flow from the heart to the lungs. The right lower heart chamber is small and underdeveloped. A life-threatening complication of tricuspid atresia is a lack of oxygen to the body's tissues. This condition is called hypoxemia.

Prompt treatment greatly improves the outcome for babies with tricuspid atresia. But complications may develop later in life. Complications of tricuspid atresia may include:

* Easy tiring during activity
* Irregular heart rhythms
* Kidney or liver disease
* Heart failure

**Prevention**

Because the exact cause of most congenital heart defects is unknown, it may not be possible to prevent tricuspid atresia.

If you have a family history of congenital heart defects or are at high risk of giving birth to a child with one, genetic screening may be recommended before or during pregnancy. Consider talking with a genetic counselor and a pediatric heart doctor about your specific risks.

Some ways you can help prevent your baby's overall risk of congenital heart defects are to:

* **Get proper prenatal care.** Regular checkups with a health care provider during pregnancy can help keep you and your baby healthy.
* **Take a multivitamin with folic acid.** Taking 400 micrograms of folic acid daily has been shown to reduce problems with the brain and spinal cord at birth. It may help reduce the risk of congenital heart defects as well.
* **Get a rubella (German measles) vaccine.** A rubella infection during pregnancy may affect a baby's heart development. Get vaccinated before trying to get pregnant.
* **Check with your provider before taking any medications.** Some medications taken during pregnancy can cause health problems in the baby. Tell your provider about all the medications you take, including those bought without a prescription.
* **Don't smoke tobacco or drink alcohol during pregnancy.** Either can increase the risk of congenital heart defects.
* **Avoid chemical exposure whenever possible.** While you're pregnant, it's best to stay away from chemicals, including cleaning products and paint, as much as you can.
* **Manage other health conditions.** If you have other health conditions, talk to your health care provider about the best way to treat and manage them.

**When to see a doctor**

Serious congenital heart defects are diagnosed before or soon after your child is born. If you notice that your baby has changes in skin color, trouble breathing, slow growth or poor weight gain, contact your healthcare provider.

## What to expect long-term

## Infants who have these surgeries are not cured; they might have lifelong complications. If the tricuspid atresia is very complex, or the heart becomes weak after the surgeries, a heart transplant might be needed. Children who receive a heart transplant will need to take medicines for the rest of their lives. The medicines help prevent rejection of the new heart.

## Babies born with tricuspid atresia will need regular follow-up visits with a cardiologist (a heart doctor). These visits can help monitor their progress and check for other health conditions that might develop as they get older. As adults, they may need more surgery or medical care for other possible problems.

## **Epidemiology**

## United States data

## An estimated 404 babies are born with tricuspid atresia each year in the United States.

## Race-, sex-, and age-related demographics

## No racial predilection is apparent.

## Considering all forms of tricuspid atresia, no sexual predilection exists. Males present more frequently with transposed great vessels than females.

## The anomaly is congenital and is evident at birth.

## **Differential Diagnoses**

* Atrial Septal Defect
* Pulmonic Stenosis (Pulmonary Stenosis)
* Tricuspid Stenosis

REFERENCES

[Tricuspid Atresia Differential Diagnoses](https://emedicine.medscape.com/article/158359-differential?form=fpf)

[About Tricuspid Atresia | Congenital Heart Defects (CHDs) | CDC](https://www.cdc.gov/heart-defects/about/tricuspid-atresia.html)

[Tricuspid atresia - Symptoms & causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/tricuspid-atresia/symptoms-causes/syc-20368392)

[Tricuspid atresia - Diagnosis & treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/tricuspid-atresia/diagnosis-treatment/drc-20368416)

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## **Total anomalous pulmonary venous return (TAPVR)**

Total anomalous pulmonary venous return (TAPVR) is a rare heart condition that's present at birth. That means it's a congenital heart defect.

Other names for this condition are:

* Total anomalous pulmonary venous connection.
* TAPVC.

In this heart condition, the lung blood vessels, called the pulmonary veins, attach to the wrong place in the heart.

In a typical heart, oxygen-rich blood flows from the lungs to the upper left heart chamber, called the left atrium. Blood then goes to the rest of the body.

In TAPVR, the connection of veins is changed. Blood flows through the upper right heart chamber, called the right atrium. This change in blood flow causes oxygen-poor blood to mix with oxygen-rich blood. As a result, blood flowing to the body doesn't have enough oxygen.

The type of TAPVR depends on where the veins connect. Most children born with TAPVR have no family history of congenital heart disease.

#### **TAPVR types**

TAPVR types differ by how blood reaches the wrong place — your baby’s right atrium. Typically, your lungs send oxygen-rich blood to your heart’s left atrium (the chamber in your heart’s upper left side). In babies with TAPVR, the oxygen-rich blood flows through pulmonary veins to their heart’s right atrium instead.

In the right atrium, the oxygen-rich blood mixes with blood that doesn’t have as much oxygen. This low-oxygen blood travels out of the heart to the rest of your baby’s body.

The TAPVR types are:

* Supracardiac TAPVR: Blood flows via an ascending vertical vein into the brachiocephalic vein and then through their superior vena cava (a large vein in their body).
* Cardiac TAPVR: Blood moves through their coronary sinus. This vein normally drains blood that contains low levels of oxygen.
* Infracardiac TAPVR: The connecting vertical vein comes from their liver and inferior vena cava.
* Mixed TAPVR: A combination of veins from your baby’s lungs drain to more than one of the types mentioned above. This is the rarest form and the most difficult one to repair successfully.

### **Symptoms**

### **What are the symptoms of total anomalous pulmonary venous return?**

Symptoms of TAPVR usually appear very soon after birth. Babies have symptoms at birth if they have any narrowing where their lung veins connect to their hearts. But some babies don’t have symptoms for several weeks.

Symptoms may include:

* Cyanosis (bluish or grayish skin, nails and lips).
* Difficulty feeding.
* Fatigue and lethargy (lack of energy).
* Heart murmur.
* Shortness of breath (dyspnea) and trouble breathing.
* Lack of growth or weight gain.

### **Causes**

Most congenital heart defects are caused by changes that happen early as the unborn baby's heart is developing before birth. An unborn baby is also called a fetus. The exact cause of most congenital heart defects, including total anomalous pulmonary venous return (TAPVR), is not known.

Changes in the genes, some medicines or health conditions, and environmental or lifestyle factors, such as smoking, may play a role.

### **Risk factors**

Possible risk factors for congenital heart defects, including total anomalous pulmonary venous return (TAPVR), may include:

* **Rubella, also called German measles.** Having rubella during pregnancy can change how the baby's heart develops. A blood test can be done before pregnancy to see if you're immune to rubella. If you're not, you can get a vaccine.
* **Diabetes.** Having type 1 or type 2 diabetes during pregnancy may change how the baby's heart grows during pregnancy. Diabetes that develops during pregnancy is called gestational diabetes. Gestational diabetes generally doesn't increase the risk of congenital heart disease.
* **Genetics.** Although TAPVR doesn't usually run in families, changes in genes have been linked to heart conditions at birth. For example, people with Down syndrome are often born with heart conditions.
* **Smoking.** If you smoke, quit. Smoking during pregnancy or exposure to secondhand smoke increases the risk of congenital heart defects in the baby.
* **Alcohol use.** Drinking alcohol during pregnancy has been linked to heart conditions in the baby.
* **Some medicines.** Some medicines taken during pregnancy may increase the risk of congenital heart defects. These include lithium (Lithobid) for bipolar disorder and isotretinoin (Claravis, Myorisan, others), which is used to treat acne. Talk to your healthcare team about the medicines you take.

### **Diagnosis**

Cardiologists can sometimes diagnose this condition with an echocardiogram before birth. Some babies don’t get a diagnosis until they’re several weeks or months old.

After your baby is born, their provider will do a physical exam and listen to your baby’s heart. A pulse oximeter (pulse ox) can measure the amount of oxygen in your baby’s blood. It fits over your baby’s big toe and sends information through a wire.

To see pictures of your child’s heart, a provider may use:

* Chest X-ray.
* CT (computed tomography) scan.
* Heart MRI (magnetic resonance imaging).
* Echocardiogram.
* Electrocardiogram (EKG or ECG).

Some of these tests (MRI) may require anesthesia, but they’re all noninvasive. They allow your baby’s healthcare provider to see images of your baby’s heart and veins. They also help them evaluate blood flow and look for abnormalities.

A provider can use cardiac catheterization to make a diagnosis. But they can usually get the information they need from imaging. This spares your baby from an invasive procedure.

### **Treatment**

Total anomalous pulmonary venous return (TAPVR) is treated with surgery. The surgery usually is done when a child is a baby. The timing of surgery depends on whether there's a blockage. To repair the heart, surgeons connect the pulmonary veins to the left upper heart chamber. They also close the hole between the upper heart chambers.

A person with TAPVR needs regular health checkups for life to check for infection, blockages or irregular heartbeats. A doctor trained in congenital heart diseases should provide care. This type of healthcare professional is called a congenital cardiologist.

### **How is total anomalous pulmonary venous return treated?**

Nearly every baby with total anomalous pulmonary venous return needs surgery to survive. Healthcare providers treat TAPVR with open-heart surgery. Most often, providers perform this surgery as soon as they can after diagnosing the condition.

Though it’s extremely rare, some people don’t have surgery as babies and get a TAPVR diagnosis as adults. But they usually have high pressures within their lungs (pulmonary hypertension) that can make surgical repair very challenging.

While waiting for surgery, your child may receive extra oxygen or a ventilator to help them breathe. They may receive an inotrope, which is a medicine that helps their heart beat more forcefully.

While your baby is asleep under general anesthesia, a surgeon:

1. Make cuts (incisions) in your baby’s chest and heart.
2. Connect the pulmonary veins to the correct place (the left atrium) in your baby’s heart.
3. Closes abnormal connections.
4. Removes any blockage in your child’s pulmonary vein.
5. Most often, it closes the hole between their left and right atria (atrial septal defect).

## **Outlook / Prognosis**

### **What can I expect if my child has total anomalous pulmonary venous return?**

Without surgery, some forms of total anomalous pulmonary venous return are typically fatal a few weeks after birth. With early diagnosis and surgical treatment, the outlook for babies with TAPVR is very good. The survival rate after surgery is around 97%.

Your child will need regular visits with their cardiologist as they grow into adulthood. Lifelong follow-up visits can help cardiologists find problems like an irregular heartbeat or blockages (obstructions) in their blood vessels. An obstruction requires another surgery and may be hard to treat.

Your child may need to take medicine or have a procedure like a cardiac catheterization.

## **Living With**

### **How do I take care of my child?**

Children with TAPVR may have trouble with typical development like fine motor function. You can help your child by getting screenings for developmental milestones. If a screening shows a developmental issue, you can arrange for someone to work with them to improve their skills.

Talk to your child’s healthcare provider about their ability to participate in sports and physical activities. Your child may need to limit vigorous exercise.

### **When should my child see their healthcare provider?**

After surgery, your baby will have checkups every six to 12 months. They’ll need regular follow-up appointments through adulthood. Your child’s provider may want to order tests like an electrocardiogram, exercise stress test or echocardiogram.

#### **When should I go to the ER?**

Get medical help right away if your baby:

* Has bluish or grayish skin, nails or lips.
* Has trouble breathing or difficulty eating.
* Seems lethargic.

### Occurrence

About 1 in every 7,809 babies in the United States are born with TAPVR[1]. This means that about 504 babies are born with TAPVR each year.

REFERENCES

<https://www.cdc.gov/heart-defects/about/tapvr.html>

[Total Anomalous Pulmonary Venous Return (TAPVR)](https://my.clevelandclinic.org/health/diseases/23069-total-anomalous-pulmonary-venous-return)

[Total anomalous pulmonary venous return (TAPVR) - Overview - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/total-anomalous-pulmonary-venous-return/cdc-20385613)

**Truncus arteriosus**

Truncus arteriosus (common truncus) occurs when a single common blood vessel comes from the heart, instead of the usual two vessels. A baby with this condition may need surgery or other procedures soon after birth. Therefore, this defect is considered a critical congenital heart defect (critical CHD).

Truncus arteriosus happens when the blood vessel from the heart in the developing baby fails to separate completely during development. This leaves a connection between the aorta and pulmonary artery. There are several different types of truncus, depending on how the arteries remain connected. There is also usually a hole between the bottom two chambers of the heart (ventricles) called a ventricular septal defect (VSD).

As blood flows to the lungs and the rest of the body, oxygen-poor blood and oxygen-rich blood are mixed together. As a result, too much blood goes to the lungs. This causes the heart to work harder to pump blood to the rest of the body.

Instead of having an aortic valve and a pulmonary valve, babies with this condition have a single common valve (truncal valve). This truncal valve controls blood flow out of the heart and is often abnormal. For example, the valve can be thickened and narrowed, which can block the blood as it leaves the heart. It can also leak, causing blood that leaves the heart to leak back into the heart across the valve.

## **Signs and symptoms**

## Infants with truncus arteriosus usually are in distress in the first few days of life. This is because of the high amount of blood going to the lungs which makes the heart work harder. Infants with truncus arteriosus can have a bluish looking skin color, called cyanosis, because their blood doesn't carry enough oxygen. Infants with truncus arteriosus or other conditions causing cyanosis can have symptoms such as:

* Problems breathing
* Fast heart rate
* Weak pulse
* Ashen or bluish skin color
* Poor feeding
* Extreme sleepiness

**Risk factors**

The exact cause of truncus arteriosus is unknown. But some things might increase the risk of a heart problem at birth. Risk factors include:

* **Viral illness during pregnancy.** Some infections can hurt a developing baby. For example, having German measles during pregnancy can cause changes in a baby's heart development. German measles also is called rubella.
* **Poorly controlled diabetes during pregnancy.** Careful control of your blood sugar before and during pregnancy can reduce the risk of heart problems in your baby. If you have diabetes, work with your healthcare professional to be sure blood sugar is well controlled before getting pregnant.
* **Some medicines taken during pregnancy.** Some medicines can cause heart problems and other health conditions in a baby. Tell your healthcare professional about all the medicines you take, including those bought without a prescription.
* **Some chromosomal disorders.** An extra or irregular chromosome increases the risk of truncus arteriosus. Examples are DiGeorge syndrome, also called 22q11.2 deletion syndrome, and velocardiofacial syndrome.
* **Smoking during pregnancy.** If you smoke, quit. Smoking during pregnancy increases the risk of heart conditions in your baby.
* **Alcohol use.** Drinking alcohol during pregnancy increases the risk of heart conditions and other health issues in a baby.
* **Obesity.** Obesity increases the risk of giving birth to a baby with a heart condition.

**Causes**

Truncus arteriosus occurs as a baby's heart forms during pregnancy. There's often no clear cause. Genetics and environmental factors may play a role.

### **How the heart works**

To understand more about truncus arteriosus, it may be helpful to know how the heart typically works.

The typical heart has four chambers. They are:

* **The right upper chamber, also called the right atrium.** This heart chamber receives oxygen-poor blood from the body.
* **The right lower chamber, also called the right ventricle.** This heart chamber pumps blood into the lungs through a large vessel called the pulmonary artery. The blood flows through the pulmonary artery into smaller vessels in the lungs where blood picks up oxygen.
* **The left upper chamber, also called the left atrium.** This heart chamber receives oxygen-rich blood from the lungs through vessels called pulmonary veins.
* **The left lower chamber, also called the left ventricle.** This chamber pumps the oxygen-rich blood to the body through the body's largest blood vessel, called the aorta.

### **A baby's heart before birth**

The way the unborn baby's heart forms during pregnancy is complex. At some point, there is a single large blood vessel leading out of the heart. The vessel is called the truncus arteriosus. It usually splits in two as the unborn baby grows in the womb. One part becomes the lower end of the body's main artery, called the aorta. The other part becomes the lower part of the pulmonary artery.

But in some babies, the truncus arteriosus never splits. The wall separating the two lower heart chambers hasn't closed all the way. This results in a large hole between those chambers, called a ventricular septal defect.

Babies with truncus arteriosus also often have a problem with the heart valve that controls blood flow from the lower heart chambers to the single vessel. This valve may not close all the way when the heart relaxes. Blood can move the wrong way, back into the heart. This is called truncal valve regurgitation.

**Diagnosis**

Truncus arteriosus is usually diagnosed soon after a child is born. The baby may look blue or gray and have trouble breathing.

When a baby is born, a healthcare professional always listens to the baby's lungs to check breathing. If a baby has truncus arteriosus, the healthcare professional may hear fluid in the lungs during this exam. The healthcare professional also listens to the baby's heart to check for irregular heartbeats or a whooshing sound, called a murmur.

**Tests**

Tests to diagnose truncus arteriosus include:

## Pulse oximetry. A sensor placed on the fingertip records the amount of oxygen in the blood. Too little oxygen may be a sign of a heart or lung problem.

## Chest X-ray. This test shows the condition of the heart and lungs. It can show the size of the heart. A chest X-ray also can tell if the lungs have extra fluid.

## Echocardiogram. An echocardiogram uses sound waves to create pictures of the beating heart. This is the main test to diagnose truncus arteriosus. It shows blood flow through the heart and heart valves. In a baby with truncus arteriosus, the test shows one single large vessel leading from the heart. There's typically a hole in the wall between the lower heart chambers.

## **Treatment**

## Infants with truncus arteriosus need surgery to improve blood flow and oxygen levels. Many procedures or surgeries might be needed, especially as a child grows. Medicines might be given before surgery to help improve heart health.

## Children and adults with surgically repaired truncus arteriosus need regular health checkups for life.

## Medications

## Some of the medicines that might be given before truncus arteriosus surgery include:

## **Wate**r pills. Also called diuretics, these medicines help the kidneys remove extra fluid from the body. Fluid buildup is a common symptom of heart failure.

## Positive inotropes. These medicines help the heart pump stronger, which improves blood flow. They also help control blood pressure. Positive inotropes may be given by IV to treat severe heart failure symptoms.

### Surgery or other procedures

## Most infants with truncus arteriosus have surgery within the first few weeks after birth. The specific type of surgery depends on the baby's condition. Usually, the baby's surgeon:

## Rebuilds the single large vessel and aorta to create a new, complete aorta.

## Separates the upper part of the pulmonary artery from the single large vessel.

## Uses a patch to close the hole between the two lower heart chambers.

## Places a tube and valve to connect the right lower heart chamber with the upper pulmonary artery. This creates a new, complete pulmonary artery.

## The tube used to create the new pulmonary artery doesn't grow with a child. Follow-up surgeries are needed to replace the tube as the child grows.

## Future surgeries may be done with a flexible tube called a catheter. This avoids the need for open-heart surgery. The healthcare professional inserts the catheter into a blood vessel in the groin and guides it to the heart. A new valve can be delivered through the catheter to the proper area.

## Sometimes a small balloon at the tip of the catheter is inflated at the site of a blockage, making a blocked artery wider. This procedure is called balloon angioplasty.

## After surgery for truncus arteriosus, a person needs lifelong follow-up care with a heart doctor specializing in congenital disease. This type of healthcare professional is called a congenital cardiologist.

## **Complications**

Truncus arteriosus causes severe problems with how blood flows through the lungs, heart and rest of the body.

Complications of truncus arteriosus in babies include:

* **Breathing problems.** Extra fluid and blood in the lungs can make it difficult to breathe.
* **High blood pressure in the lungs, also called pulmonary hypertension.** This condition causes the blood vessels in the lungs to narrow. It becomes hard for the heart to pump blood into the lungs.
* **Enlargement of the heart.** Pulmonary hypertension and increased blood flow strain the heart. The heart must work harder to pump blood. This causes the heart muscle to grow larger. The enlarged heart gradually weakens.
* **Heart failure.** In this condition, the heart cannot supply the body with enough blood. Too little oxygen and too much strain on the heart can lead to heart failure.

Infants who had their hearts successfully fixed with surgery may still have complications later in life. Possible complications are:

* Pulmonary hypertension that gets worse.
* Backward flow of blood through a heart valve, called regurgitation.
* Irregular heartbeats, called arrhythmias.

Common symptoms of these complications include:

* Dizziness.
* Feeling very fast, fluttering heartbeats.
* Feeling very tired.
* Shortness of breath when exercising.
* Swelling of the belly, legs or feet.

### **Truncus arteriosus in adults**

In rare cases, some people born with truncus arteriosus can survive without heart surgery. They may live into adulthood. But those with the condition will almost certainly have heart failure and develop a complication called Eisenmenger syndrome. This syndrome is caused by permanent lung vessel damage. It results in a significant lack of blood flow to the lungs.

## **Prevention**

Because the cause is unclear, it may not be possible to prevent truncus arteriosus. Getting good prenatal care is important. If you or someone in your family had a heart condition present at birth, talk to your healthcare professional before getting pregnant. You might need to see a genetic counselor and a heart doctor, called a cardiologist.

If you decide to get pregnant, taking these steps can help keep your baby healthy:

* **Get recommended vaccinations.** Some infections can be harmful to a developing baby. For example, having German measles — also called rubella —during pregnancy can cause changes in a baby's heart development. A blood test done before pregnancy can show if you're immune to rubella. A vaccine is available for those who aren't immune.
* **Talk to your healthcare professional about your medicines.** Check with your healthcare professional before taking any medicines if you're pregnant or thinking about getting pregnant. Many drugs aren't recommended for use during pregnancy because they can harm a developing baby.
* **Take a folic acid supplement.** Take a multivitamin with folic acid. Taking 400 micrograms of folic acid daily has been shown to reduce brain and spinal cord conditions in babies. It may help reduce the risk of heart conditions present at birth too.
* **Control diabetes.** If you have diabetes, ask your healthcare professional how to best manage the disease during pregnancy.

**Lifestyle and home remedies**

If you or your child had truncus arteriosus, your healthcare professional may recommend taking a few steps to protect the heart.

* **Exercise limits.** Some people with heart conditions need to limit exercise and sports activities, especially competitive sports. Ask your healthcare professional which sports and types of exercise are safe for you or your child. People with Eisenmenger syndrome should avoid strenuous physical activity.
* **Antibiotics.** Sometimes, heart conditions can increase the risk of infection in the lining of the heart or heart valves. This infection is called infective endocarditis. Antibiotics may be recommended to prevent infections before dental procedures and other surgeries. It's also important to have good oral hygiene and regular dental checkups
* **Pregnancy.** If you've had truncus arteriosus repair surgery and want to become pregnant, talk to your healthcare professional first. Ask about the possible risks and complications. It's best to be checked by healthcare professionals with training in adult congenital heart disease and high-risk pregnancies. Together, you and your care team can discuss and plan for any special care needed during pregnancy.  
  Depending on the level of lung damage that occurred before truncus arteriosus surgery, pregnancy might or might not be recommended. Pregnancy is considered a very high risk for those with Eisenmenger syndrome and is not recommended.

**EPIDEMIOLOGY**

### Occurrence

Truncus arteriosus occurs in 1 out of every 15,984 live births. It can occur by itself or as part of certain genetic disorders. There are about 230 cases of truncus arteriosus per year in the United States.

**DIFFERENTIAL DIAGNOSIS**

New technological advances have improved the rate of diagnosing truncus arteriosus prenatally to greater than 90%. However, truncus arteriosus may be misdiagnosed as pulmonary atresia with a VSD if structures and aortopulmonary collaterals cannot be correctly identified on prenatal echocardiography. Other differential diagnoses for a single outflow tract ventricle include a double outlet right ventricle with pulmonary and aortic atresia.

REFERENCES

[Truncus Arteriosus - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK534774/#article-30641.s4)

[Truncus arteriosus - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/truncus-arteriosus/diagnosis-treatment/drc-20364277)

[Truncus arteriosus - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/truncus-arteriosus/symptoms-causes/syc-20364247)

[About Truncus Arteriosus | Congenital Heart Defects (CHDs) | CDC](https://www.cdc.gov/heart-defects/about/truncus-arteriosus.html)

**Aortic coarctation**

Coarctation of the aorta occurs when a baby's aorta doesn't form correctly as the baby develops during pregnancy. The aorta is a tube that carries oxygen-rich blood from the heart to the rest of the body. Babies with this condition have a part of their aorta that is narrower than usual.

The narrowing, or coarctation, blocks normal blood flow to the body. This can back up flow into the left ventricle of the heart. As a result, the muscles in this ventricle work harder to get blood out of the heart.

If the narrowing is severe enough and it is not diagnosed, the baby may have serious problems soon after birth. For this reason, coarctation of the aorta is considered a critical congenital heart defect (critical CHD).

Aortic coarctation (ko-ahrk-TAY-shun) is a narrowing of a part of the body's main artery, called the aorta. The condition forces the heart to work harder to pump blood.

Coarctation of the aorta is usually present at birth. That means it is a congenital heart defect. But sometimes the condition can occur later in life.

Coarctation of the aorta often occurs along with other congenital heart defects. Treatment to fix the condition is usually successful. But regular health checkups are needed for life to watch for changes in the heart's health.

**Causes**

The cause of coarctation of the aorta is unclear. It's usually a heart problem present at birth, called a congenital heart defect. A congenital heart defect happens as the baby is growing in the womb during pregnancy. The cause is often unknown.

Rarely, coarctation of the aorta can happen later in life. Conditions or events that can narrow the aorta and cause this condition include:

* Traumatic injury.
* An extreme buildup of cholesterols and fats in the arteries, called atherosclerosis.
* A rare type of swelling and irritation of blood vessels in the heart, called Takayasu arteritis.

## Signs and symptoms

## The narrowing of the aorta is usually located after arteries branch to the upper body. In infants, coarctation in this region can lead to:

* Normal or high blood pressure and pulsing of blood in the head and arms
* Low blood pressure and weak pulses in the legs and lower body

In babies with a more severe form of coarctation of the aorta, early signs may include:

* Pale skin
* Irritability
* Heavy sweating
* Difficulty breathing

Older children and adults with coarctation of the aorta often have high blood **pressure in the arms.**

**COMPLICATIONS**

Long-term high blood pressure. Blood pressure usually drops after surgery to fix the aorta. But it may still be higher than usual.

* A weakened or bulging artery in the brain, also known as a brain aneurysm.
* Bleeding in the brain.
* A rupture or tear in the body's main artery, called an aortic dissection.
* A bulge in the wall of the body's main artery, called an aortic aneurysm.
* Coronary artery disease.
* Stroke.

Prompt treatment is needed to help prevent complications. Without treatment, coarctation of the aorta may lead to:

* Kidney failure.
* Heart failure.
* Death.

Some people have complications after treatment for coarctation of the aorta. These complications include:

* Re-narrowing of the aorta, called re-coarctation.
* Aortic aneurysm or rupture.

To prevent complications, people with coarctation of the aorta need regular health checkups for life.

**RISK FACTORS**

Risk factors for coarctation of the aorta include:

* Male sex.
* Some genetic conditions, such as Turner syndrome.
* Some heart conditions present at birth, called congenital heart defects.

Congenital heart defects associated with coarctation of the aorta include:

* **Bicuspid aortic valve.** The aortic valve is between the body's main artery and the lower left heart chamber. If the aortic valve has only two flaps, called cusps, instead of the usual three, it's called a bicuspid valve.
* **Subaortic stenosis.** This is a narrowing of the area below the aortic valve. It blocks blood flow from the lower left heart chamber to the aorta.
* **Patent ductus arteriosus.** The ductus arteriosus is a blood vessel that connects the left lung artery to the aorta. When a baby is growing in the womb, this vessel lets blood go around the lungs. Shortly after birth, the ductus arteriosus usually closes. If it stays open, the opening is called a patent ductus arteriosus.
* **Holes in the heart.** Some people with coarctation of the aorta also are born with a hole in the heart. If the hole is between the upper heart chambers, it is called an atrial septal defect. A hole between the lower heart chambers is called a ventricular septal defect.
* **Congenital mitral valve stenosis.** This is a type of heart valve disease that some people are born with. The valve between the upper and lower left heart chambers is narrowed. It's harder for blood to move through the valve.

## Diagnosis

## Coarctation of the aorta is usually diagnosed after the baby is born. How early in life the defect is diagnosed usually depends on how mild or severe the symptoms are.

## Coarctation of the aorta is a critical CHD. However, newborn screening with pulse oximetry might be less likely to detect this condition.

## Detection of the defect is often made during a physical exam. The pulse will be noticeably weaker in the legs or groin than it is in the arms or neck. A heart murmur—an abnormal whooshing sound caused by disrupted blood flow—may be heard through a doctor’s stethoscope. Older children and adults with coarctation of the aorta often have high blood pressure in the arms.



A heart murmur may be heard through a doctor's stethoscope.

Once suspected, the healthcare provider might request one or more tests to confirm the diagnosis. The most common test is an echocardiogram, which is an ultrasound of the heart. It will show the location and severity of the coarctation and whether any other heart defects are present. Other tests to measure the function of the heart may be used including:

* Chest x-ray
* Electrocardiogram (EKG)
* Magnetic resonance imaging (MRI)
* Cardiac catheterization

## **Echocardiogram.** Sound waves are used to create images of the beating heart. An echocardiogram shows how blood flows through the heart. The test can often tell which part and how much of the aorta is narrowed. An echocardiogram also helps healthcare professionals plan treatment for coarctation of the aorta.

## **Electrocardiogram (ECG or EKG).** This quick and painless test checks the heartbeat. Sensors called electrodes stick to the chest and sometimes to the arms or legs. Wires connect the sensors to a machine, which displays or prints results. If the aorta is very narrowed, an ECG may show of thickening of the walls of the lower heart chambers.

## **Chest X-ray.** A chest X-ray shows the condition of the heart and lungs. A chest X-ray might show a narrowing in the aorta at the site of the coarctation.

## **Cardiac magnetic resonance imaging (MRI).** This test uses magnetic fields and radio waves to create detailed images of the heart and blood vessels. It can show which part and how much of the aorta is narrowed. A healthcare professional also may use MRI results to guide treatment.

## **Computerized tomography (CT) scan.** A CT scan uses a series of X-rays to create detailed cross-sectional images of the body.

## **Coronary angiogram with cardiac catheterization.** A coronary angiogram uses X-rays to look at the heart's blood vessels, called the coronary arteries. It's usually done to see if a blood vessel is narrowed or blocked. A coronary angiogram is part of a general group of heart tests and treatments called cardiac catheterization. During cardiac catheterization, a thin flexible tube called a catheter is placed in a blood vessel, usually in the groin or wrist, and guided to the heart. Dye flows through the catheter to arteries in the heart. The dye makes the arteries easier to see on X-ray images and video. Cardiac catheterization can help determine how much of the aorta is narrowed.

## **CT angiogram.** This test looks at the arteries that supply blood to the heart. It uses a powerful X-ray machine to make images of the heart and its blood vessels. A CT angiogram uses a dye and special X-rays to show how blood flows through the veins and arteries. The test can show the location and severity of coarctation of the aorta. It also can tell whether other blood vessels are affected. A CT angiogram also can be used to guide treatment.

## Treatment and recovery

## **Surgery or other procedures**

## Surgery or a heart procedure can be done to repair aortic coarctation. Options include:

* **Balloon angioplasty and stenting.** This may be the first treatment for aortic coarctation. Sometimes it's done if the aorta gets narrow again after coarctation surgery. The treatment helps widen a narrowed artery and improve blood flow.  
  During angioplasty, a doctor uses a thin tube called a catheter and a tiny balloon to open a narrowed artery. Usually, a small metal coil called a stent is placed in the artery. The stent keeps the artery open. It also reduces the risk of renarrowing.
* **Resection with end-to-end anastomosis.** A surgeon removes the narrowed area of the aorta. This is called a resection. The surgeon then connects the two healthy parts of the aorta. This is called anastomosis.
* **Subclavian flap aortoplasty.** A surgeon takes part of the blood vessel that delivers blood to the left arm, called the left subclavian artery, and uses it to widen the narrowed area of the aorta.
* **Bypass graft repair.** This surgery uses a tube called a graft to create a new path for blood to flow around the narrowed area of the aorta.
* **Patch aortoplasty.** The surgeon cuts across the narrowed area of the aorta and patches in a piece of material to expand the blood vessel. This treatment is useful if the coarctation involves a long part of the aorta.

No matter what age the coarctation is diagnosed, the narrow aorta will need to be widened once symptoms are present. This can be done with a procedure called balloon angioplasty or through surgery.

A balloon angioplasty uses a thin, flexible tube, called a catheter. The catheter is inserted into a blood vessel and directed to the aorta. When the catheter reaches the narrow area of the aorta, a balloon at the tip is inflated. This helps to expand the blood vessels.

Sometimes a mesh-covered tube (stent) is inserted to keep the vessel open. The stent is often used to initially widen the aorta or re-widen it if the aorta narrows again after surgery.

During surgery to correct a coarctation, the narrow portion is removed. The aorta is reconstructed or patched to allow blood to flow normally through the aorta.

**Lifestyle and home remedies**

People who have coarctation of the aorta need to take steps to control blood pressure and watch for complications. Follow these tips:

* **Get regular exercise.** Regular exercise helps lower blood pressure. Talk with your healthcare team about the amount and type of exercise that's best for you. Some physical activities, such as weight lifting, can temporarily raise blood pressure.
* **Talk with your healthcare team before getting pregnant.** Coarctation of the aorta, even after it's fixed, may increase the risk of an aortic tear or rupture during pregnancy and childbirth. Before becoming pregnant, talk with a doctor trained in congenital heart diseases, called an adult congenital cardiologist. Together you can go over the possible risks and complications.
* **Prevent heart infections.** Bacteria can affect the inner lining of the heart or valves, causing an infection called endocarditis. People with some heart conditions may need to take antibiotics before dental work to prevent this infection. Ask your healthcare professional whether you need preventive antibiotics.

**EPIDEMIOLOGY**

Coarctation of the aorta is a relatively common congenital heart defect, accounting for approximately 5% to 7% of all congenital heart diseases and occurring more frequently in males, with a male-to-female ratio of about 2:1. Coarctation of the aorta imposes an increased afterload on the left ventricle (LV), often leading to LV hypertrophy and, ultimately, heart failure if left untreated. In the United States, LV diastolic dysfunction is responsible for over half of heart failure admissions, with heart failure being the leading cause of death among adults with congenital heart disease, contributing to around 45% of cardiovascular-related deaths.

Coarctation of the aorta is associated with other forms of congenital heart disease, such as a bicuspid aortic valve, Shone complex, and an interrupted aortic arch. A bicuspid aortic valve occurs in 45% to 75% of patients with coarctation of the aorta. The classic Shone complex comprises a specific set of congenital heart defects, including aortic coarctation, subaortic membrane, parachute mitral valve, and a supravalvar mitral ring; this precise combination is uncommon. More frequently, patients exhibit a series of left-sided obstructions involving one or more of these lesions, often accompanied by other mitral valve abnormalities or hypoplasia of the LV.

Critical coarctation of the aorta is a severe form seen in neonates where adequate blood flow to the lower body relies on blood from the pulmonary artery through the patent ductus arteriosus into the descending aorta. A similar condition is an interrupted aortic arch, characterized by a complete discontinuity between the aortic arch and descending aorta. The location of this interruption often determines the presence of associated defects, such as an aortopulmonary window or a ventricular septal defect, found in up to 75% of cases.

The epidemiology of aortic coarctation also reveals a significant genetic component. Turner syndrome is strongly associated with coarctation of the aorta, leading to an increased risk of left-sided obstructive heart lesions; karyotype screening is recommended for females diagnosed with aortic coarctation. Additionally, first-degree relatives of individuals with obstructive left-sided heart lesions have a 10-fold increased risk of developing aortic coarctation and other congenital heart defects. Despite advancements in early detection and treatment, coarctation of the aorta remains a significant contributor to cardiovascular morbidity and mortality, particularly when associated with other congenital anomalies.

**DIFFERENTIAL DIAGNOSIS**

Coarctation of the aorta can present diagnostic challenges due to its variable clinical manifestations, which often overlap with other conditions, particularly those causing secondary hypertension. While this anomaly is frequently diagnosed during infancy and childhood, it can remain undetected for many years, particularly in patients who present with hypertension later in life. This overlap in clinical presentation necessitates a careful differential diagnosis that includes:

* Renal parenchymal diseases
  + Renal parenchymal diseases, such as glomerulonephritis or polycystic kidney disease, are among the most common causes of secondary hypertension in adolescents. Renal ultrasound and serum creatinine levels are essential investigations when renal pathology is suspected.
* Renovascular disease
  + Renovascular hypertension, caused by conditions such as renal artery stenosis, is another key cause of secondary hypertension in this age group. The incidence of renovascular disease is approximately 6.69 per 100,000 person-years, with a lower incidence in patients younger than 18. Doppler ultrasonography, CTA, or MR angiography (MRA) can help identify renal artery stenosis.
* Primary aldosteronism
  + This condition involves excessive production of aldosterone, leading to hypertension and hypokalemia. Diagnosis is obtained by measuring plasma aldosterone concentration and plasma renin activity measurements.
* Pheochromocytoma
  + A catecholamine-secreting tumor can cause paroxysmal or sustained hypertension, often accompanied by headaches, sweating, and palpitations. Diagnosis is made through urinary or plasma metanephrines.
* Cushing disease and syndrome
  + This condition is characterized by hypercortisolemia, leading to hypertension, obesity, and glucose intolerance. Diagnosis involves the dexamethasone suppression test and measurement of 24-hour urinary-free cortisol.
* Other cardiovascular anomalies
  + Other congenital heart diseases, such as aortic stenosis, could also present with similar symptoms, including hypertension and diminished lower extremity pulses.

**Importance of Considering Coarctation of the Aorta**

Given the significant overlap of symptoms, particularly hypertension, aortic coarctation should be considered a priority in the differential diagnosis, especially in adolescents presenting with secondary hypertension. This particular congenital anomaly remains a significant but often underrecognized cause of hypertension, mainly because of its variable presentation and the common association of hypertension with more prevalent renal or endocrine disorders. Given the reported prevalence of congenital heart disease, it is vital to include coarctation in the differential diagnosis when evaluating an adolescent with unexplained hypertension.

When suspected, coarctation of the aorta is best evaluated with imaging studies such as echocardiography, which can identify the characteristic narrowing of the aorta and assess for associated cardiac anomalies. CTA or MRA can also provide detailed anatomical information and guide therapeutic decisions. Early diagnosis and appropriate management are crucial for preventing the long-term complications of untreated coarctations, such as severe hypertension, heart failure, and aortic dissection.

REFERENCES

[Coarctation of the Aorta - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK430913/#article-37463.s4)

[Coarctation of the aorta - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/coarctation-of-the-aorta/diagnosis-treatment/drc-20352535)

[About Coarctation of the Aorta | Congenital Heart Defects (CHDs) | CDC](https://www.cdc.gov/heart-defects/about/coarctation-of-the-aorta.html)

[Coarctation of the aorta - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/coarctation-of-the-aorta/symptoms-causes/syc-20352529)

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## **Double-outlet right ventricle**

Double-outlet right ventricle is a heart condition present at birth. That means it's a congenital heart defect. In this condition, the lower right heart chamber has two openings for blood to exit the heart instead of one. Some of the blood from the lower right heart chamber goes to the body instead of the lungs.

Babies with double-outlet right ventricles also have a hole between the lower heart chambers. The lower heart chambers are called the ventricles. The hole is called a ventricular septal defect. The hole causes oxygen-rich blood to mix with oxygen-poor blood. Babies with this hole may not get enough oxygen in the bloodstream. Their skin may appear gray or blue.

Double-outlet right ventricle may occur with other heart conditions present at birth such as other holes in the heart and blood vessels or heart valve changes.

### **Symptoms**

Symptoms of double-outlet right ventricle may include:

* Pale gray or blue lips, tongue, or fingernails. Depending on skin color, these changes may be harder or easier to see.
* Rapid breathing.
* Problems with feeding, growth and weight gain.

#### **When to see a doctor**

If you think that your baby has symptoms of a heart condition, make an appointment for a health checkup right away.

### **Causes**

The exact cause of a double-outlet right ventricle is not known. The condition happens when the unborn baby, also called a fetus, is developing before birth.

Gene changes, some medicines or health conditions, and environmental or lifestyle factors may play a role. Smoking while pregnant also may increase the risk of certain congenital heart defects in an unborn baby.

Usually, the body's main artery, called the aorta, connects to the left lower heart chamber. The pulmonary artery connects to the right lower heart chamber. But in babies with double-outlet right ventricle, both the aorta and the pulmonary artery connect partially or completely to the right lower heart chamber.

### **Risk factors**

Having a family history of congenital heart conditions may increase the risk of congenital heart defects such as double-outlet right ventricle.

### **Complications**

Complications of double-outlet right ventricle include poor growth and heart failure.

### **Diagnosis**

A test called an echocardiogram can diagnose the double-outlet right ventricle. The test uses sound waves to create images of the beating heart. It can show blood flow through the heart and heart valves.

If more details about the heart are needed, other tests may be done. Tests to check the heart may include:

* **Heart CT scan.** This also is called a cardiac CT. This test uses X-rays to create cross-sectional images of specific parts of the body.
* **Heart MRI scan.** This test uses magnetic fields and radio waves to create detailed pictures of the heart.
* **Cardiac catheterization.** A doctor places a long, thin flexible tube called a catheter into a blood vessel, usually in the groin or wrist. It's guided to the heart. Dye flows through the catheter to the heart arteries. The dye helps the arteries show up more clearly on images taken during the test. During this test, the healthcare professional also can measure pressure and oxygen levels in the chambers of the heart and in the blood vessels.

### **Treatment**

Most babies with double-outlet right ventricles have heart repair surgery within a few months of age. More than one type of surgery may be done. The type of surgery depends on the specific heart structure.

The heart surgeon may do one or more of the following to treat double-outlet right ventricle:

* Create a pathway in the heart to connect the lower left heart chamber to the body's main artery, called the aorta.
* Use a patch, called a baffle, to fix the hole in the heart.
* Fix the aorta and pulmonary artery positions if they're reversed.
* Connect the lower right heart chamber to the pulmonary artery, if necessary.
* Widen the natural pathway to the lungs if it's narrowed.

Other surgeries might be needed later in life. A person born with double-outlet right ventricle needs regular health checkups for life. Adults should see a doctor trained in evaluating and treating congenital heart conditions. This type of doctor is called an adult congenital cardiologist

## **Diagnostic Considerations**

## **Other problems to consider**

Distinguish double outlet right ventricle (DORV), with or without transposition of the great arteries, and subaortic ventricular septal defect (VSD) from isolated VSD.

DORV with pulmonary valve stenosis (PS) may have a presentation similar to that of tetralogy of Fallot (TOF).

DORV with subpulmonary VSD but without PS may have a presentation similar to that of transposition of the great arteries with VSD.

## Important considerations

Medicolegal pitfalls in caring for patients with double outlet right ventricle and transposition of the great arteries are similar to those for any patient with congenital heart disease (CHD).

Failure to make the correct diagnosis is of paramount importance. The correct treatment plan can be determined only if all anatomic details are known. Misdiagnosis can lead to inappropriate care.

Another issue is surgery. Because most of these patients do well when medical/surgical care is administered at a center with considerable experience in caring for infants with CHD, referral to such a center provides the best opportunity for a good long-term outcome.

The physician must be familiar with the possible complications that may result from surgery and be able to treat complications resulting from surgery appropriately.

Prenatal diagnosis can be made with detail in most cases of DORV

**Epidemiology**

Double outlet right ventricle (DORV) accounts for about 2-3% of all congenital heart defects, with a birth prevalence rate of 1/ 10,000.

REFERENCE.

[Double-outlet right ventricle - Overview - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/double-outlet-right-ventricle/cdc-20389537)

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# **Single Ventricle Defects**

## **DEFINITION**

Rare disorders affecting one lower chamber of the heart. The chamber may be smaller, underdeveloped, or missing a valve.

Hypoplastic Left Heart Syndrome (HLHS) - An underdeveloped left side of the heart. The aorta and left ventricle are too small and the holes in the artery and septum did not properly mature and close.

Pulmonary Atresia/Intact Ventricular Septum - The pulmonary valve does not exist, and the only blood receiving oxygen is the blood that is diverted to the lungs through openings that normally close during development.

Tricuspid Atresia - There is no tricuspid valve in the heart so blood cannot flow from the body into the heart in the normal way. The blood is not being properly refilled with oxygen so it does not complete the normal cycle of body–heart–lungs–heart–body.

### Hypoplastic Left Heart Syndrome

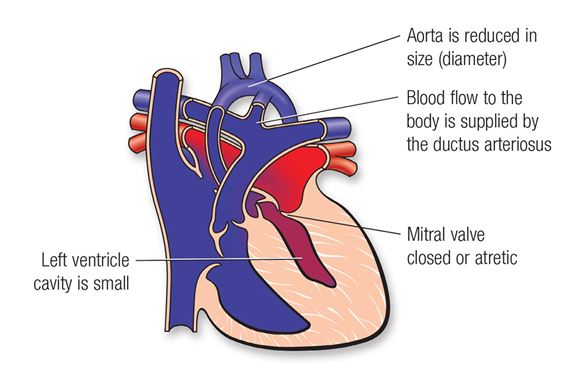
In hypoplastic left heart syndrome (HLHS), the heart's left side — including the aorta, aortic valve, left ventricle and mitral valve — is underdeveloped.

#### What causes it?

In most children, the cause isn't known. Some children can have other heart defects along with HLHS.

#### How does it affect the heart?

In HLHS, blood returning from the lungs must flow through an opening in the wall between the atria (atrial septal defect). The right ventricle pumps the blood into the pulmonary artery and blood reaches the aorta through a patent ductus arteriosus (see diagram).



#### How does the defect affect my child?

The baby often seems normal at birth but comes to medical attention within a few days of birth as the ductus closes. The baby may appear ashen, have rapid and difficult breathing and have difficulty feeding. This heart defect is usually fatal within the first days or month of life unless it's treated.

#### What can be done about the defect?

Although this defect isn't correctable, some babies can be treated with a series of operations, or heart transplantation. Until an operation is performed, the ductus arteriosus is kept open by intravenous medication. Because these operations are complex and need to be adapted for each child, it's necessary to discuss all the medical and surgical options with your child's doctor.

If you and your child's doctor agree that surgery should be performed, it will be done in several stages. The first stage (Norwood procedure) allows the right ventricle to pump blood to both the lungs and the body without the need for the ductus to be kept open. Blood is directed to the lungs through either a Blalock-Taussig (arrow on image below) or Sano shunt. The Norwood procedure must be performed soon after birth. The second stage (bidirectional Glenn or hemi-Fontan) is usually performed between 4 and 8 months and the third stage (lateral tunnel Fontan or extracardiac Fontan) is usually performed between 18 months and 3 years.

These operations create a connection between the veins returning low-oxygen blood to the heart and the pulmonary artery. The goal is to allow the right ventricle to pump only oxygenated blood to the body and to prevent or reduce cyanosis (lower than normal blood oxygen levels). Some infants require several intermediate operations to achieve this.

Some doctors recommend heart transplantation to treat HLHS. Although it can provide the infant with a heart that has normal structure, the infant will require life-long medications to prevent rejection. Many other transplant-related problems can develop, and these should be discussed with your child's doctor.

#### What activities can my child do?

Children with HLHS may be advised to limit their physical activities to their own endurance. Generally, many competitive sports pose greater risk. Your child's pediatric cardiologist will help determine the proper level of activity.

#### What will my child need in the future?

Children with HLHS require lifelong follow-up by a cardiologist for repeated checks of how their heart is working. Virtually all children with HLHS will require heart medicines, heart catheterization and additional surgery.

#### What about preventing endocarditis?

Children with HLHS are at increased risk for developing endocarditis. Ask your pediatric cardiologist about your child's need to take antibiotics before certain dental procedures to help prevent endocarditis.

**EPIDEMIOLOGY**

The occurrence of congenital heart disease is between 6 and 13 in 1,000 live births. HLHS, the most common form of univentricular heart disease, is seen in 2 to 3 per 10,000 births, with a higher incidence in male infants.Tricuspid atresia occurs in about 1 per 10,000 live births. Ebstein anomaly is observed in about 0.5 per 10,000 live births with sex predilection. However, with maternal use of lithium, the likelihood of Ebstein anomaly can increase nearly 7 times. Double outlet right ventricle occurs in 0.009 cases per 10,000 live births. Double inlet left ventricle is reported in up to 0.01 per 10,000 live births. Atrioventricular canal defect is found in 0.03 to 0.04 per 10,000 live births.

**DIFFERENTIAL DIAGNOSIS**

Neonatal symptoms may appear immediately after birth or within a few days as collateral circulations diminish. In infants with severe manifestations after delivery, the differential diagnoses should include sepsis, infantile respiratory distress syndrome, aortic or pulmonary disorders, and transposition of the great arteries. Milder cases may not present until months or years later, detected only when growth is impaired. In such cases, infection, failure to thrive, and malnutrition should be investigated as potential causes.

REFERENCES

[Single Ventricle - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK557789/#article-81926.s4)

[Single Ventricle Defects | American Heart Association](https://www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/single-ventricle-defects)