Empower | Understand | Thrive

EMPOWER MY CONGENITAL HEART

The Shape of My Heart

Understanding Your Anatomy, Repairs, and Future

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CHD OVERVIEW

How common is CHD?

- **1 in 100** people are born with CHD.
- It is estimated that **there are more adults than children** living with CHD.
- In the U.S., it is estimated that **2 million adults** are living with CHD, the **number increasing by 40 - 50K every year.**

Types of CHD

There are many types of CHDs, ranging from simple to complex. A person may have **one defect or a combination** of several. A combination of CHDs can occur in two ways:

- **Multiple separate defects**: A person may have more than one defect, such as a ventricular septal defect (VSD) and a coarctation of the aorta, or a VSD and pulmonary stenosis.
- **A condition made up of several defects**: Some CHDs are defined by a specific set of defects that occur together. For example, Tetralogy of Fallot is one complex condition that includes four defects: a VSD, pulmonary stenosis, a thickened right ventricle, and a misaligned aorta.

MANAGING YOUR CHD

The type of repair you have—or may need—depends on **when** you were born (since repair methods have changed over time), the **experience** of the hospital and care team performing the repair, and the **age** at which it was done.

Even after repair, your heart **requires lifelong checkups** to ensure all its components — the structure, the pump, the electrical system, and the plumbing — continue to function correctly. Think of it like maintaining a house: regular care helps prevent bigger problems later.

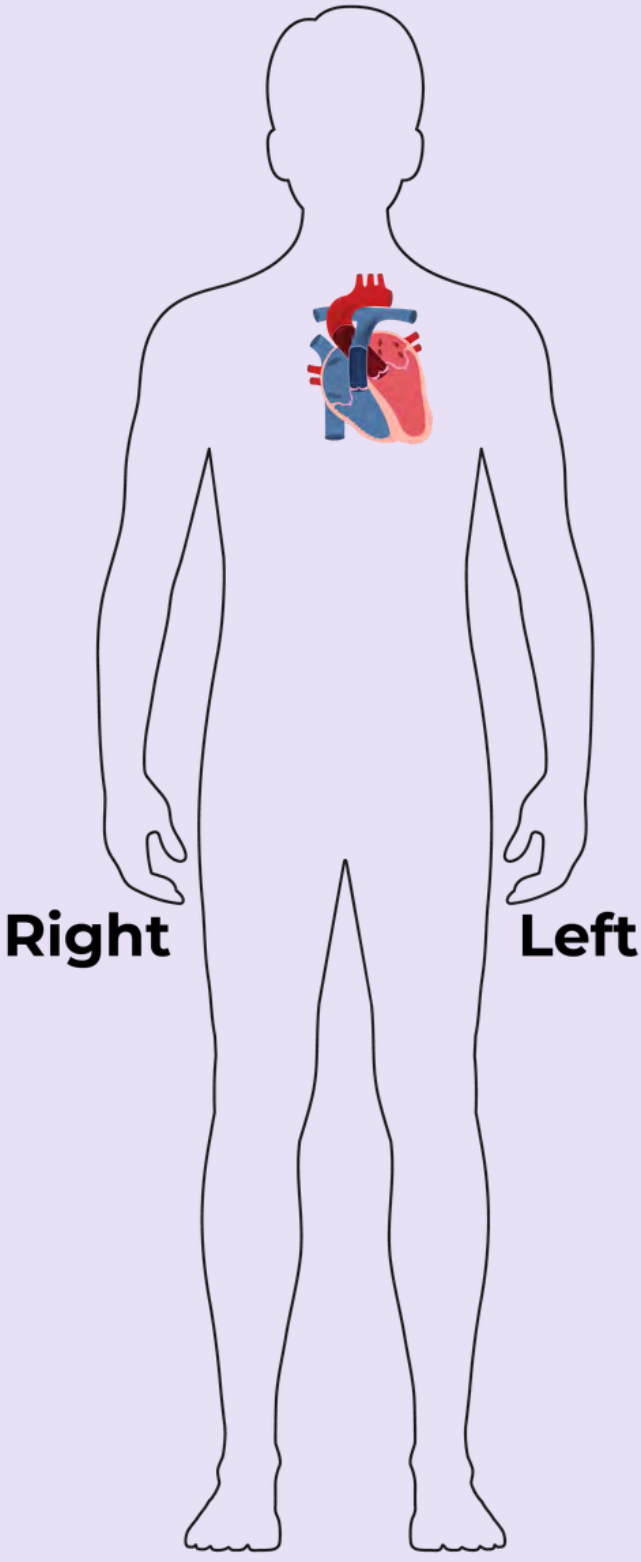
All these factors underscore why you should **see an ACHD cardiologist for your lifelong care,** whenever possible. Most providers, including general cardiologists, may not fully understand the nuances of these different CHDs and their repairs in adult patients.

Research shows that by **regularly** seeing an **ACHD cardiologist**, you are more likely to:

- **Live Longer!**
- **Live Healthier!**
- **Avoid Complications!**

If you don't have access to an ACHD cardiologist, consider having your local cardiologist coordinate with one.

NORMAL ORIENTATION OF THE HEART IN THE BODY



NORMAL BLOOD FLOW

By understanding how blood flows through a normal heart, you'll be better prepared to understand how your CHD may change that.

Right Side (Pulmonary Circulation):

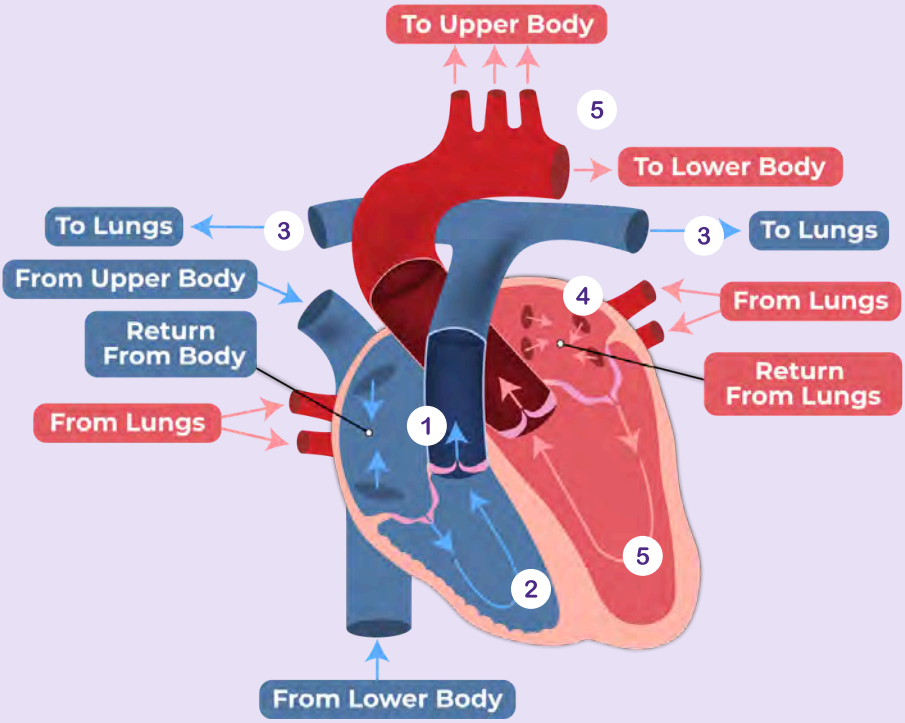
1. Blood that has ***delivered oxygen*** to the body returns to the right atrium.
2. It moves into the right ventricle, which pumps it through the pulmonary artery (plumbing) to the lungs.
3. In the lungs, blood picks up oxygen.

This side works as a low-pressure system.

Left Side (Systemic Circulation):

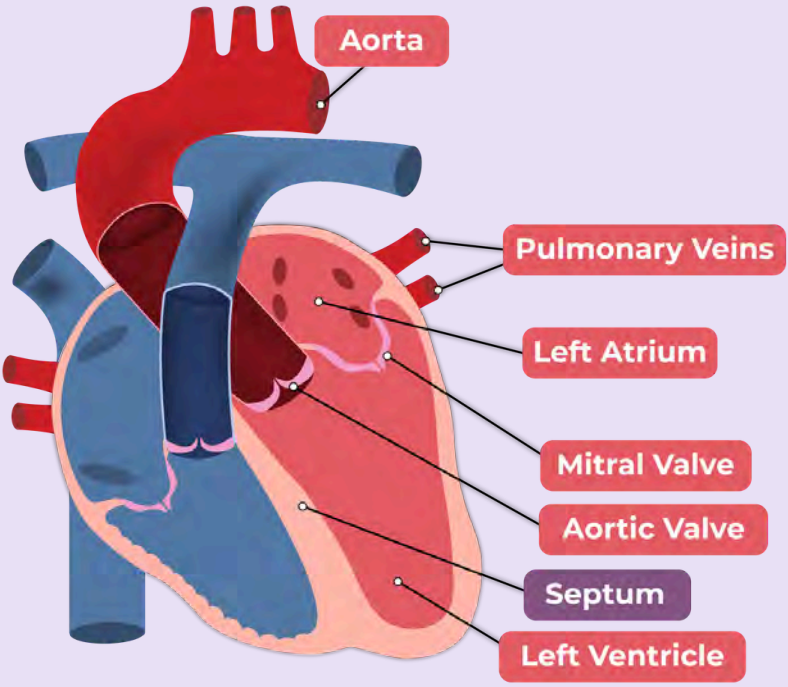
4. ***Freshly oxygenated*** blood returns from the lungs to the left atrium.
5. It moves into the left ventricle, which pumps it out through the aorta (plumbing) to the body.

This side works as a high-pressure system.

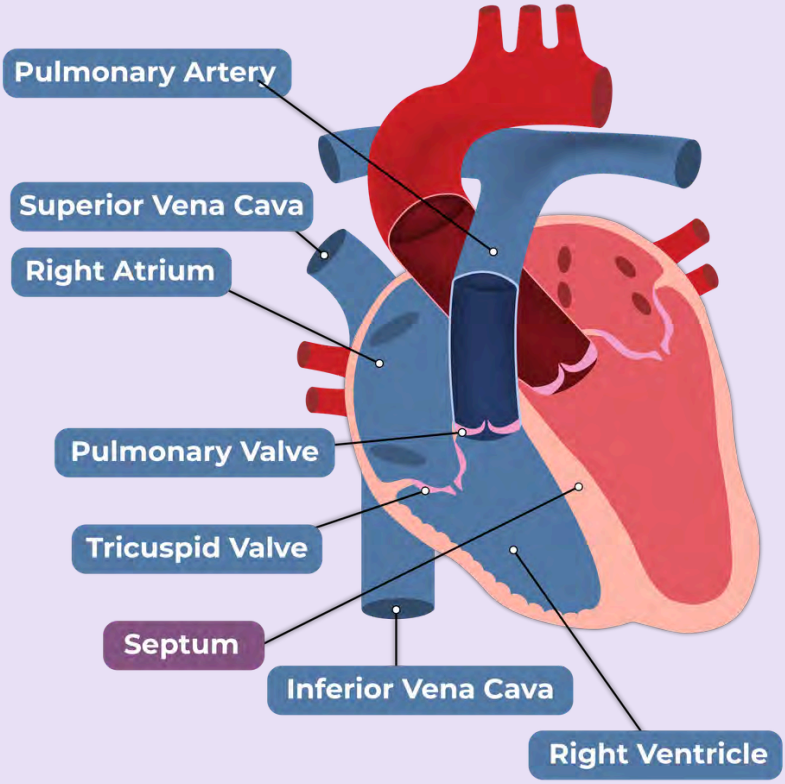


NORMAL ANATOMY

The ***left side*** is a high-pressure system designed to pump blood throughout the entire body.



The ***right side*** is a low-pressure system, only needing enough force to push blood into the nearby lungs.

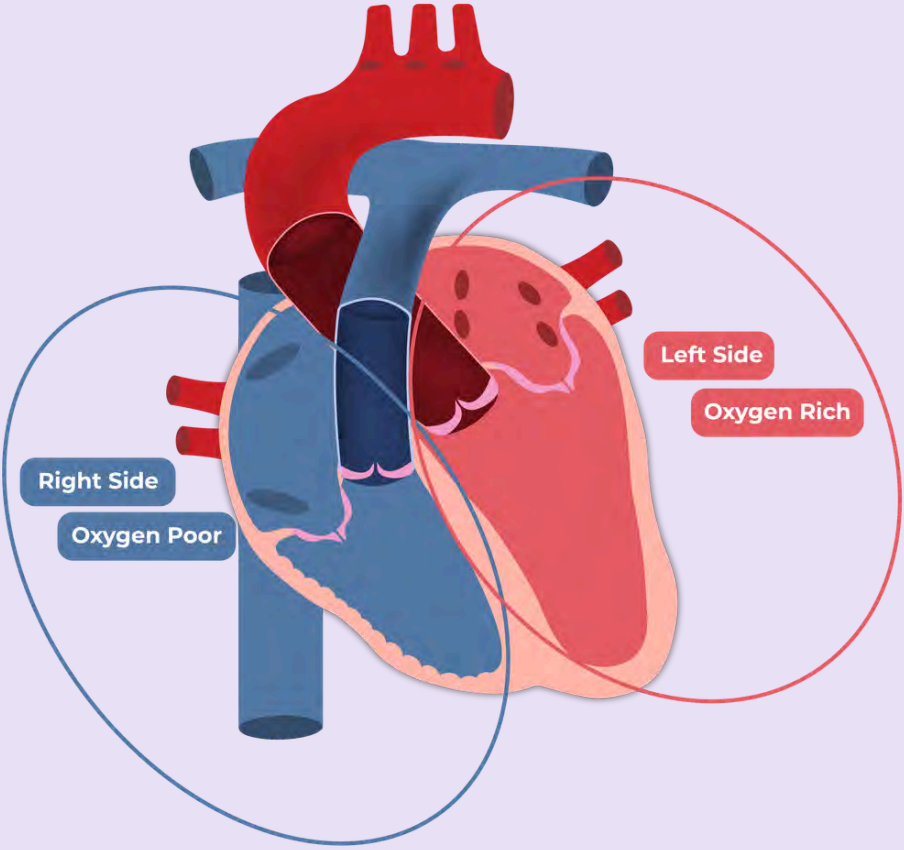


MAINTAINING SEPARATE CIRCULATIONS

One of the heart's main structural roles is to keep **oxygenated** and **less oxygenated** blood separate. Keeping these two circulations separate ensures that:

- The body always receives **oxygenated** blood.
- The ***lungs are protected*** **from the high pressures** needed to supply the whole body with blood.

Like a house with separate pipes for clean water and wastewater, the heart's structure keeps oxygenated blood separate from less oxygenated blood so they don't mix.



COMMON WAYS THE HEART CAN BE BUILT OR FUNCTION DIFFERENTLY

Valve Regurgitation / Insufficiency (Leaking)

What regurgitation means: A valve does not close tightly, allowing some blood to flow back into the chamber (atria or ventricle) that it came from.

How it affects blood flow: The heart must pump more to move the same amount of forward-flowing blood.

Possible impact: Over time, this can lead to fatigue, shortness of breath, or swelling.

Stenosis & Atresia

What stenosis means: A part of the heart (such as a valve or blood vessel) is narrowed, making it harder for blood to flow through.

What atresia means: A part of the heart did not form fully and is either closed or missing, blocking normal blood flow.

Possible impact: The heart must work harder to pump blood through the narrowing or around the blockage, which can cause symptoms such as fatigue or shortness of breath.

COMMON WAYS THE HEART CAN BE BUILT OR FUNCTION DIFFERENTLY

Shunts

What a shunt is: A hole or extra connection between the left and right sides of the heart allows blood to flow differently than normal.

How it affects blood flow: Because pressure is different on each side, oxygen-rich and oxygen-poor blood can mix.

Possible impact: A shunt you are born with can cause the heart to work harder to pump blood correctly. This can lead to tiredness, shortness of breath, or poor growth.

Did You Know?

A shunt can be created as part of a surgical repair **or** could be part of the heart defect you are born with.

Dilation

What dilation means: A heart chamber becomes stretched or enlarged.

How it affects blood flow: If the heart is dilated, the walls can weaken over time, reducing the heart's ability to squeeze properly.

Possible impact: This can cause fluid buildup, swelling, or shortness of breath.

COMMON WAYS THE HEART CAN BE BUILT OR FUNCTION DIFFERENTLY

Decreased Pumping Function

What it means: The heart muscle becomes weaker and cannot push enough blood forward with each beat.

How it affects blood flow: Blood may back up into the lungs or body, leading to fluid buildup and swelling.

Possible impact: People may feel tired, short of breath, or notice swelling in their legs or abdomen.

Hypertrophy & Hypoplasia

What hypertrophy means: A heart chamber or muscle becomes abnormally thick because it has been working harder than normal—**hypertrophic** is the term used to describe a structure affected by hypertrophy.

What hypoplasia means: A heart chamber, valve, or vessel is underdeveloped and smaller than normal, which limits how much blood it can handle—**hypoplastic** is the term used to describe a structure affected by hypoplasia.

Possible impact: Both can reduce efficient blood flow and increase strain on the heart.

COMMON REPAIR DEVICES

What are patches?

A patch is a small piece of material placed during surgery to close a hole, widen a narrowed area, or rebuild part of the heart.

What are occluders?

An occluder is a small device placed through a catheter to plug a hole without open-heart surgery.

What are balloons?

A balloon is inflated during a catheter procedure to stretch a narrow valve or blood vessel and improve blood flow.

What are stents?

A stent is a small mesh tube left inside a vessel after ballooning to keep the pathway open.

What are conduits?

A conduit or homograft is a tube that replaces or creates a missing connection between the heart and major blood vessels, directing blood where it needs to go when the natural pathway is too small or absent.

What are coils?

A spiral coil is placed inside extra blood vessels (collaterals) to block the abnormal blood flow.

COMMON PROCEDURES

Valve Repairs & Replacements

What valve repair means: The original valve is repaired so it can open and close more normally, often by removing extra tissue or adding support.

What valve replacement means: The damaged valve is removed and replaced with a new one, which may be made of tissue or mechanical materials.

How they help: These procedures improve blood flow and reduce strain on the heart.

Cardioversion & Ablation

What cardioversion does: If you develop a fast or irregular heart rhythm (such as atrial flutter, atrial fibrillation, or ventricular tachycardia), a brief electrical shock may be delivered to reset the heart back to a normal rhythm.

What ablation does: Either hot or cold energy is applied to stop irregular heartbeats (arrhythmia) and restore normal rhythm. This can usually be done with a catheter procedure, but sometimes it is done during surgery if you need surgery for another reason.

INFECTION, ANTIBIOTICS, & CHD

People with CHD can have a **risk of infection of the heart** structures (endocarditis).

One way to prevent infection is with antibiotic prophylaxis, which means taking an **antibiotic before certain procedures.**

When taking antibiotics, be aware that **unnecessary antibiotics can hide early signs of a serious infection** and may affect blood culture results, making diagnosis more challenging.

Always check with your heart doctor before starting any antibiotics.

EMPOWERMENT

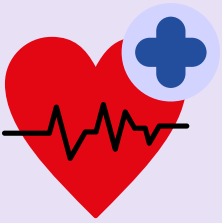
If you're prescribed antibiotics to treat a fever, you can insist on a blood culture **BEFORE** taking them. This ensures doctors can identify the specific bacteria if you have an infection.

INFECTION, ANTIBIOTICS, & CHD

Reduce Infection Risk with Dental Care

- **Brush and floss** your teeth twice a day.
- Schedule **cleanings every 6 months.**
- It is common for people with CHD to take **antibiotics before a dental procedure** to *prevent* infection.
- Tell your dental team about your heart condition, **confirm the antibiotic is recommended** for you, and take it exactly as prescribed.

If you have fevers, chills, or night sweats after your appointment, call your doctor ***immediately***.



HEART FAILURE

What is heart failure, really?

Heart failure means the heart is **not pumping as well as it should**, not that it is about to stop.

It **can be temporary**, especially when caused by an infection, high blood pressure, valve problems, or a heart rhythm issue.

With the right treatment, heart function often improves.

How is it diagnosed?

Doctors diagnose heart failure mainly by listening to your symptoms, which might include **shortness of breath, swelling, or fatigue**, and by examining you.

Tests like echocardiograms can help, but the diagnosis is based primarily on **how you feel** and what your provider observes.

LEARN ABOUT YOUR HEART CONDITION

After this page, there is a list of the **most common CHD** diagnoses. Please select the condition(s) you'd like to learn more about today.

After selecting your condition(s), you can explore:

- What it is & what it does
- Statistics (how common it is)
- Common repairs & procedures
- Long-term implications
- Common symptoms
- Recommended follow-up

When you finish, you'll have the option to:

1. **Email yourself a full PDF** that includes all CHD conditions (not just the ones you viewed).
2. Share your **feedback** to help us improve this tool.

If you don't see your specific diagnosis listed, it may be closely related to one of the conditions shown. Please use the feedback form at the end to tell us which diagnosis you were hoping to find.

CHD CONDITIONS PAGES

WHAT ARE AORTIC VALVE CONDITIONS?

This section explains Aortic Valve conditions, including:

- **Aortic valve stenosis** (the valve is too narrow)
- **Aortic valve regurgitation** (the valve is leaky)
- **Bicuspid, unicuspid,** or **quadricuspid aortic valve**

If you have Shone's complex or Coarctation of Aorta, please review the section for those conditions.

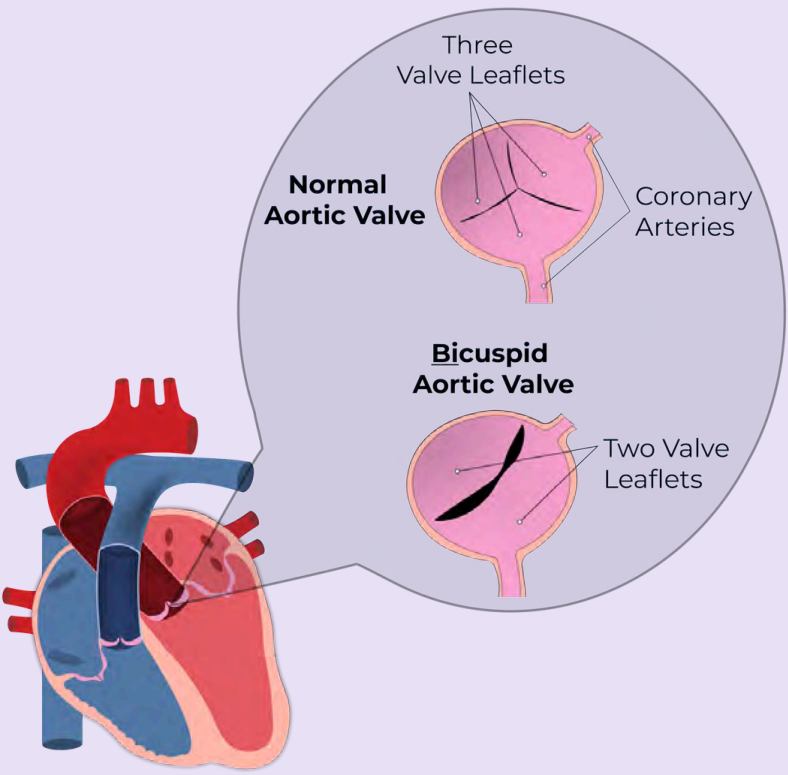
If you were born with an absent aortic valve or a very small left ventricle, please review the single ventricle section.

You will be able to receive a PDF of all the conditions at the end.

WHAT ARE AORTIC VALVE CONDITIONS?

Structural Issues

The aortic valve allows blood to flow from the heart's main pumping chamber (left ventricle) to the body's main artery (aorta). The valve usually has 3 leaflets (cusps) that open and close the valve with each heartbeat (like a door). Some people are born with a **unicuspid** (1 leaflet), **bicuspid** (2 leaflets), or **quadricuspid** (4 leaflets) aortic valve.



What Happens?

Your valve may become **narrow** (stenosis), making it harder for blood to flow out/or **leaky** (regurgitation), causing blood to flow backward.

AORTIC VALVE CONDITIONS: WHAT TO KNOW

Statistics

Aortic valve disorders account for **3 - 6%** of all CHD. Bicuspid aortic valve is the most common, occurring in **4 - 5 cases among 1000** live births

Associated With:

Structural Issues

Coarctation of aorta: Narrowing of the aorta.
Aortic dilation: Enlargement of the aorta. It may also occur specifically in the first section of the aorta (aortic root).
Shone's complex: Multilevel narrowing on the left side of the heart (at the level of the mitral valve, at or below or above the aortic valve, aortic arch).
Ventricular septal defect: Hole between the lower two heart chambers.

Genetics

Turner syndrome: There is a single X chromosome instead of two.
Familial inheritance: There is 7% risk of having a bicuspid valve among immediate relatives (parents, siblings, children)

AORTIC VALVE CONDITIONS: SURGICAL PROCEDURES

If your valve is significantly altered, you may need a procedure to repair or replace it.

Surgical Procedures

An invasive procedure performed to open the narrowing (valvotomy) or replace the valve.

Aortic valve repair: Preferred option if not severe enough to require replacement

Aortic valve replacement: If there is narrowing or leaking across the valve.

Aortic root replacement: If the aorta is significantly enlarged.

Ross procedure: Involves replacement of the aortic valve with the patient's own pulmonary valve, along with another new pulmonary valve. If the new pulmonary valve eventually becomes narrow or leaky, it might require replacement.

If you have associated conditions such as Coarctation of Aorta or Ventricular Septal Defect, they may require repair as well.

AORTIC VALVE CONDITIONS: CATH PROCEDURES & MEDICATIONS

Catheter Procedures

A less-invasive procedure where a **balloon** is inflated across the narrowed aortic valve to stretch it open.

In some cases, aortic valve replacement can also be performed via a catheter procedure (also known as transcatheter aortic valve replacement (TAVR)).

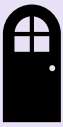
This is **most effective for valvar stenosis**

Medication Management

Used to help improve the pumping function of the left ventricle; treat high blood pressure; treat fluid buildup in the lungs or legs.

Blood thinners: If you have a mechanical aortic valve, you will be placed on blood thinners to avoid clot formation.

AORTIC VALVE CONDITIONS: LONG-TERM



***Valve Issues*:** The aortic valve can become narrow or leaky after catheter or surgical procedures, or the replaced prosthetic aortic valve may wear out over time.

Infection Risk: The aortic valve and/or heart lining may become infected (endocarditis).

Structural Issues: There is a high risk of enlargement of the aorta (dilation) or rupture (dissection) in patients with bicuspid aortic valves.

Pump Issues: The left ventricle may become thickened (hypertrophied) from pumping against the narrowed valve or enlarged from a leaky valve. Over time, this may weaken the left ventricular pumping function.

Electrical Issues: Long-standing aortic valve stenosis, scarring, or stretching of heart chambers can increase the risk of fast arrhythmias (atrial fibrillation, ventricular tachycardia). Sometimes heart rate may be too slow (bradycardia).

Not everyone will experience all long-term complications or symptoms.

AORTIC VALVE CONDITIONS: COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Chest pain, shortness of breath: Especially with exercise.

*Some people may be **asymptomatic** (do not experience any symptoms).*

Why do these things happen?

Narrowing or leaking of the aortic valve
Dilation of the aorta
Scarring in heart chambers
Decrease in the pumping function of the left ventricle

What are the potential risks?

Aortic dissection: If there is significant dilation of the aorta, there is a risk of rupture (dissection), which often shows with severe chest pain and shortness of breath. ***Dissection is an emergency!***

AORTIC VALVE CONDITIONS: FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 2 years

ECG/EKG, Echocardiogram:
every 2 years

Exercise Test, Heart Monitor:
every 2 - 3 years

Cardiac CT/MRI:
as needed

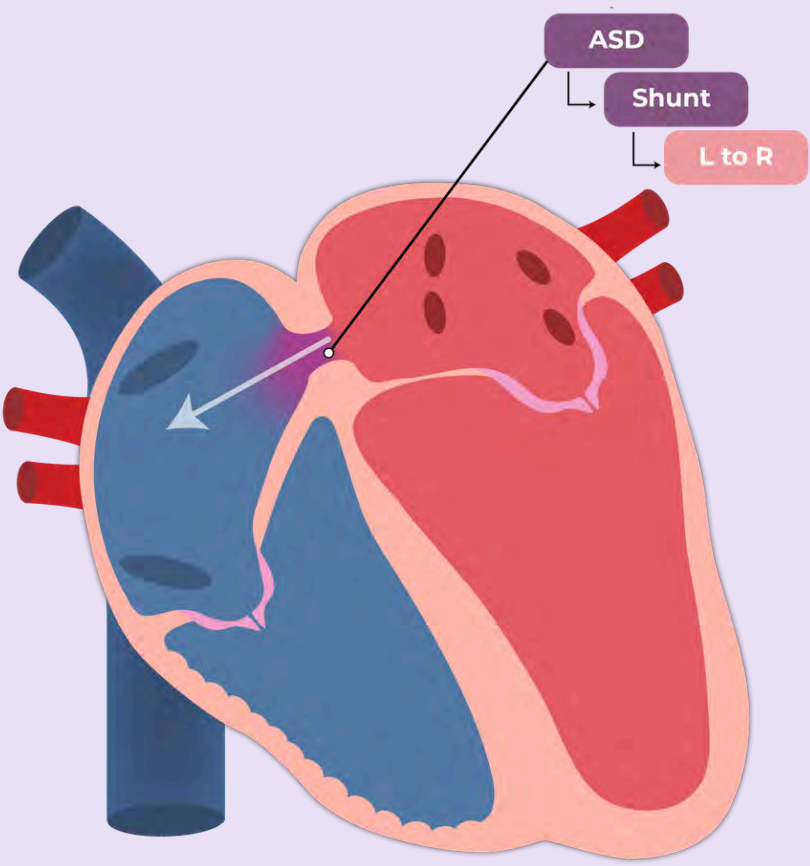
**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS AN ATRIAL SEPTAL DEFECT (ASD)?

Structural Issues

An ASD is a **hole** in the wall (septum) that separates the **two upper chambers** (right and left atria) of the heart.



What Happens?

Blood flows the **wrong way**. Some of the blood that should move to the body instead flows back across the wall (septum) between the atria into the right side of the heart. This is a left (L) to right (R) shunt.

ASD:
WHAT TO KNOW

Statistics

- ASD is a common CHD, making up **10 - 15%** of all CHDs
- ASD is the most common CHD diagnosed in adults.

Associated With:

Structural Issues

Partial Anomalous Pulmonary Venous Return (PAPVR): Some of the veins carrying blood from your lungs are connected to the wrong side of your heart, going to the right side instead of the left side.

Endocardial Cushion/Atrioventricular septal defect (AVSD): The ASD may occur with other septal defects and valve abnormalities.

If you also have PAPVR or AVSD, you should also review the sections on those defects .

You will be able to receive a PDF of all the conditions at the end.

ASD:
MANAGEMENT

Does my ASD need surgery?

ASDs occur in different parts of the septum, and the location often determines the repair type.

Small defects often don't need to be repaired if they aren't causing any problems.

Larger defects usually require **repair.** Depending on their size, location, and complexity, this may be done through a catheter procedure or surgery.

Catheter Procedure

A less invasive procedure where a device (such as a septal occluder) is placed across the hole. **Secundum ASD** is most often repaired with a catheter procedure.

Surgical Procedure

The surgeon makes an incision in the chest to directly close the hole using a tissue patch. ASDs that often require surgery include:

- **Primum ASD**: Often associated with AVSD. *(See: AVSD Section)*
- **Sinus Venosus ASD**: Often associated with PAPVR. *(See: PAPVR Section)*
- **Coronary Sinus ASD** **least common*

**A catheter procedure is preferred, if feasible*

LARGE UNREPAIRED **ASD:** LONG-TERM

Structural Issues: If the ASD is unrepaired, over time, extra blood flows from the left to the right side of the heart and then to the lungs.

Pump Issues*:* The right side of the heart must handle more blood, which can cause the heart chambers (atria or ventricles) to enlarge or weaken.

Electrical Issues: Enlarged upper chambers (atria) can lead to abnormal heart rhythms, such as atrial flutter.

Plumbing Issues: Too much blood going to the lungs can cause high pressure in the lung arteries (pulmonary hypertension). This can make the right heart work harder and sometimes cause blood to flow the wrong way (right-to-left), especially during exercise.

Valve Issues: An enlarged right ventricle can stretch the tricuspid valve, causing it to leak. In some types of ASD (like primum ASD), the nearby mitral valve can also be abnormal, leading to narrowing or leakage (*see AVSD section*).

Not everyone will experience all long-term complications or symptoms.

LARGE UNREPAIRED **ASD:** COMMON SYMPTOMS

You may not have any symptoms, and your ASD might be found by chance.

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Cyanosis: You may notice a bluish color on your lips or fingers during exercise.

Why do these things happen?

Swelling: Can happen from weak heart function, leaky valves, or high lung pressure.

Cyanosis: The blood can start flowing the opposite way across the hole in the heart wall —from the right side to the left instead of from left to right.

What are the potential risks?

Stroke: Blood clots can pass through the hole and travel to the brain.

Lung Damage: Blood pressure in your lungs can increase over time and damage the vessels.

LARGE UNREPAIRED **ASD:** TREATMENT PATHS

If Lung Pressures are Normal

- The primary treatment is to **repair the hole** (either by catheter or surgical methods).
- You might need **medication** to get rid of **excess fluid** or to strengthen your heart.
- **Rhythm problems** can be treated with medications or, rarely, an ablation.
- You may need **blood thinners** to reduce the risk of blood clots.

If Lung Pressures are High

- You may need **medications to help lower lung pressures,** such as sildenafil or macitentan.
- ASD repair is sometimes delayed until the pressure in your lungs improves.
- You may need **blood thinners** to lower your risk of stroke.
- You may need medications that help your heart pump more effectively.

LARGE UNREPAIRED ASD: FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient):
every 1-2 years

ECG/EKG, Echocardiogram:
every 1-2 years

Heart Monitor, Exercise Test:
as needed

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If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

REPAIRED **ASD:** LONG-TERM

Most people do well after the hole is repaired

Electrical Issues:

You **need long-term follow-up** since there is a risk of abnormal heart rhythm (arrhythmias) from the scarring in the atria after repair.

You may require **medications** or an **ablation** procedure to treat arrhythmias, depending on the frequency and your symptoms.

Not everyone will experience all long-term complications or symptoms.

REPAIRED ASD:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 2 - 4 years

ECG/EKG, Echocardiogram, Heart Monitor:
2 - 4 years

Exercise Test:
as needed

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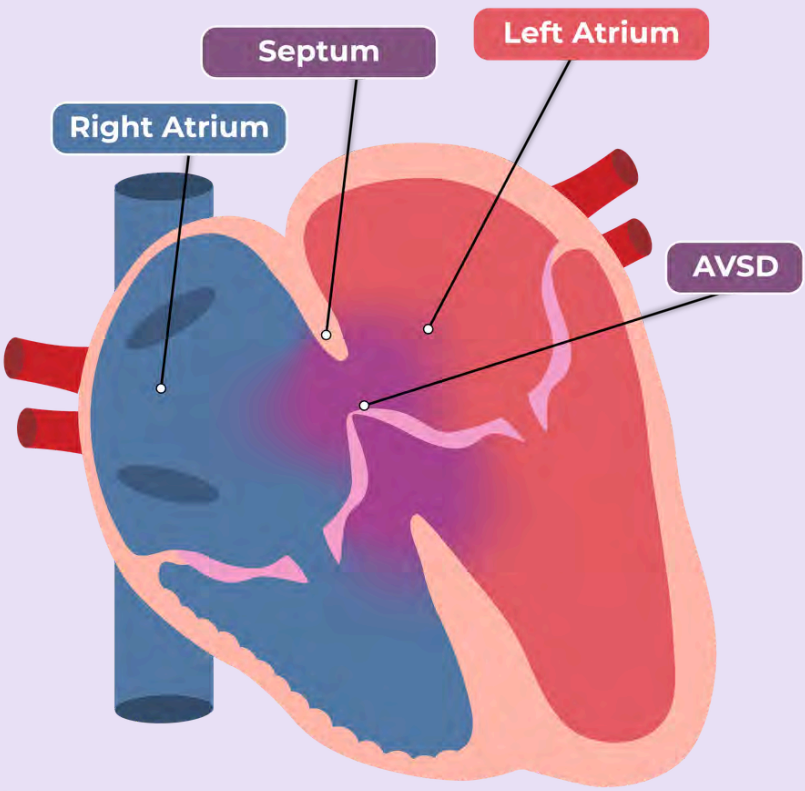
WHAT IS AN ATRIOVENTRICULAR SEPTAL DEFECT(AVSD)?

Structural

An AVSD, or **Endocardial Cushion Defect**, is a condition where there is a hole in the wall(s) (septum) that separates the two upper chambers (atria) and/or the two lower chambers (ventricles).

Valve Issues

The valves between the upper and lower chambers (mitral or tricuspid valves) do not form or work normally and can leak, causing blood to flow backward from the ventricle into the atria.



AVSD:
WHAT TO KNOW

What Happens?

Blood flows the wrong way. Some of the blood that should move to the body instead flows across the hole(s) into the right side of the heart and lungs (this is a left (L) to right (R) shunt).

Statistics

AVSD make up **4 - 5%** of all CHD

AVSD Has 2 Main Types:

Balanced AV canal: Both ventricles are well-developed and share blood flow evenly, allowing for a normal two-ventricle repair.

Unbalanced AV canal: One ventricle is smaller and receives most of the blood flow, often requiring a single ventricle surgical pathway. If you have this condition, please review the section on single ventricle.

AVSD: MANAGEMENT

Surgery is the main treatment, and is often (but not always) performed in childhood.

Surgical Procedures

During surgery, the hole(s) in the septum are covered with a **tissue patch,** and the abnormal mitral or tricuspid valves are repaired, as needed.

*If you have an AVSD that was never repaired, or if you had surgery later in adulthood, there is a chance you have developed **high lung pressure**. Your doctor may refer to this as Pulmonary Hypertension or Eisenmenger Syndrome.*

***Next Step**: Please visit the Eisenmenger Syndrome section. You will be able to receive a PDF of all the conditions at the end.*

REPAIRED **AVSD:** LONG-TERM

If you have a **repaired AVSD,** you might not have any symptoms, or you could have extra fluid in your lungs and body (symptoms related to heart failure).

Valve Issues: Over time, the mitral or tricuspid valves may leak (regurgitation) or become narrow (stenosis). This may be repaired using a catheter procedure or surgery.

Pump Issues: The atria or ventricles may become enlarged or weak. This often results from valve issues. You may need medications to help your heart's pumping function.

Electrical Issues: Due to enlarged heart chambers or scarring, there is a risk of abnormal heart rhythms, such as atrial fibrillation/atrial flutter or heart block.

Not everyone will experience all long-term complications or symptoms.

REPAIRED AVSD:
LONG-TERM

Plumbing Issues: Issues with the mitral valve can cause blood to back up into the lungs, leading to a risk of increased lung pressure (pulmonary hypertension). These include:

- **Leaky Valve** (regurgitation): If the valve leaks, blood flows backward into the lungs rather than forward. This extra fluid increases pressure in the lungs.

- **Narrowed Valve** (stenosis): If the mitral valve is too tight, blood cannot flow forward easily. This causes pressure to "back up" into the lungs, much like a clogged drain.

Infection Risk: There is an increased risk of heart infection (endocarditis) if you have a prosthetic valve or a hole that was closed with a patch.

Not everyone will experience all long-term complications or symptoms.

REPAIRED AVSD:
COMMON SYMPTOMS

Possible Symptoms

Depending on your specific heart repair and valve function, you may experience:

Tiredness: Feeling worn out faster than usual.

Swelling: Extra fluid in your legs, belly, or lungs (often caused by weak heart function or leaky valves).

Irregular heartbeats: Feeling your heart racing, fluttering, or skipping beats (palpitations).

Signs of Infection

If you develop a heart infection (endocarditis), symptoms often appear suddenly, including: flu-like symptoms, night sweats, sudden fatigue, or exercise Intolerance.

Not everyone will experience all long-term complications or symptoms.

REPAIRED AVSD:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 1 - 2 years

ECG/EKG, Echocardiogram:
every 1 - 2 years

Exercise Test, Heart Monitor:
as needed

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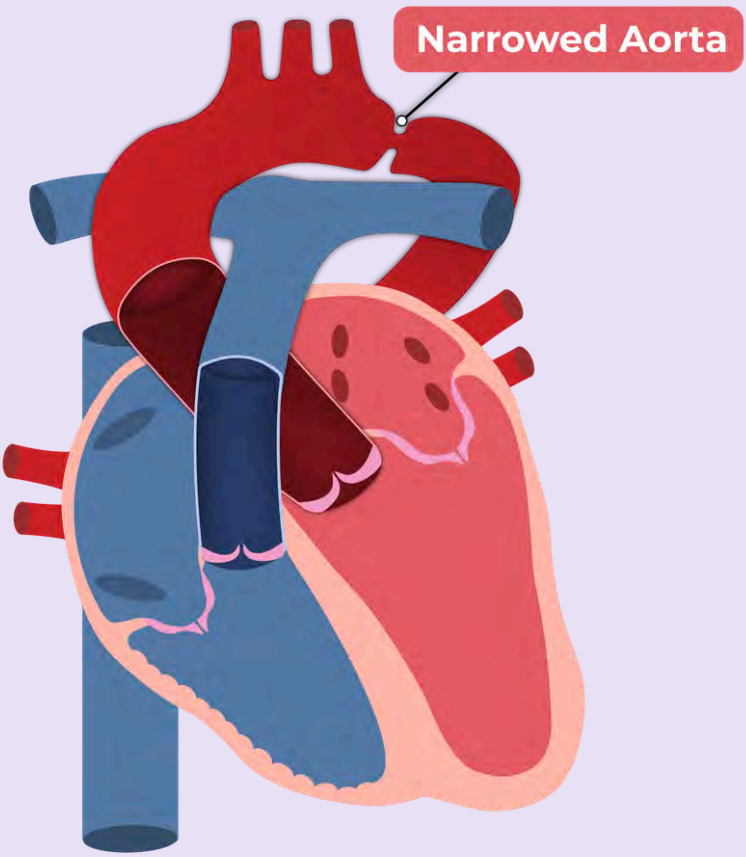
WHAT IS COARCTATION OF THE AORTA (CoA)?

Structural Issues

Narrowed Aorta: CoA is a narrowing of the aorta as it travels down to the feet, leading to decreased blood flow to the lower body.

Pump Issues

Heart Strain: Because of this narrowing, the left lower chamber (left ventricle) has to pump harder to push blood through the aorta. Since blood backs up behind the narrowed area of the aorta, blood pressure can be higher in your arms.



CoA:
WHAT TO KNOW

Statistics

5 - 8% of patients with congenital heart conditions have CoA.

Associated With:

Valve Issues

Bicuspid Aortic Valve (BAV): occurs in some patients with coarctation, where the aortic valve has two leaflets instead of the usual three.

Structural Issues

Ventricular Septal Defect (VSD): a hole in the wall (septum) between the heart's two lower chambers (ventricles).

Genetics

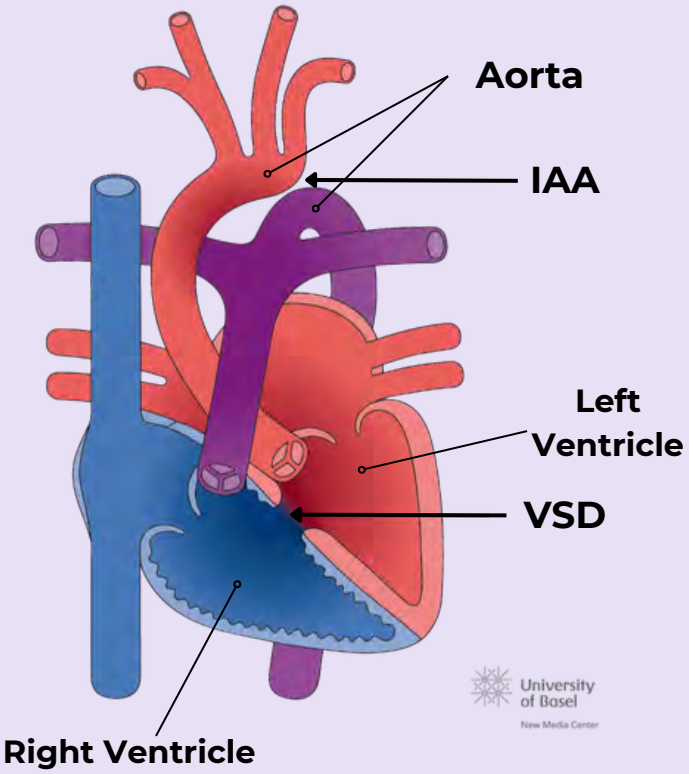
Turner Syndrome: A chromosomal condition affecting females (with a missing or incomplete X chromosome).

What about Interrupted Aortic Arch (IAA)?

Interrupted aortic arch is a **rare and severe** congenital heart defect affecting the aorta.

Structural Issues

IAA results in **severe narrowing or separation** in the aorta, creating an "interruption" in the normal arch of the aorta. Most people with an IAA also have a hole in the heart (VSD).



Statistics

IAA is rare, affecting **~1.5%** of those with CHD.

CoA / IAA: MANAGEMENT

Repair type usually depends on the age you were diagnosed and the severity of the narrowing.

Surgical Procedures

Most Common Surgical Repair: The narrowed portion of the aorta is removed, and the healthy ends are sewn back together (end-to-end anastomosis) — this surgery is common in infancy.

Other Types of Surgical Repairs:

- **A patch** can be used to make the narrowed section of the aorta larger (tissue patch or subclavian flap)
- An **artificial tube** is used to replace the narrowed section (particularly when severe or involves a long segment of the aorta). (called interposition graft).

Catheter Procedure

A **balloon catheter** procedure involves guiding a thin tube with a balloon to the narrow spot, inflating the balloon to stretch the artery open, and then placing a stent. This is most often used when a coarctation of the aorta is found **later in childhood or during adulthood**.

CoA / IAA: MANAGEMENT

What if you are diagnosed with CoA in adulthood?

You may need a repair, which is most commonly performed using a catheter procedure.

Catheter Procedures

The narrowed portion is stretched open using a **balloon**.

A **metal mesh tube** (stent) may be placed to keep the artery open.

**If a catheter procedure isn't suitable, a surgical repair may be done instead.*

Medication Management

You may need to take medication to keep your blood pressure within a safe range, even if you feel fine.

CoA / IAA:
LONG-TERM

Plumbing Issues

Recurrent Coarctation: The suture site may become narrow and not grow adequately, causing another narrowing in the aorta. This can lead to higher blood pressure in the arms than in the legs and is often treated with a catheter procedure.

Aneurysm: The aorta near the site of the repair may stretch or bulge (aortic aneurysm). About 10% of CoA patients have a small weak spot in a brain blood vessel (intracranial/brain aneurysm).

Coronary artery disease: Because of long-term high blood pressure, you may develop acquired heart conditions earlier than expected. Most commonly, this is a narrowing of the arteries supplying blood to the heart (coronary arteries).

Close monitoring and good blood pressure control are important as there is a risk that the aneurysm could burst (dissection).

Not everyone will experience all long-term complications or symptoms.

CoA / IAA:
LONG-TERM

Pump Issues: Even after a successful repair, blood pressure can remain high. This makes the left ventricle work harder to pump blood, causing the heart muscle to thicken and become stiff over time. Your blood pressure may be normal at rest, but rise abnormally with exercise.

Valve Issues: If you have a bicuspid aortic valve, there is a risk of valve narrowing (stenosis) or valve leakage (regurgitation).

Electrical Issues: Abnormal heart rhythms (arrhythmia) can sometimes occur with CoA. Pumping against the narrowing thickens and stiffens the heart muscle. This stiffness puts strain on the upper heart chamber (the left atrium), which can trigger irregular rhythms such as atrial fibrillation.

Infection Risk: There is a risk of infection in the heart (endocarditis) at the site of surgical repair, valve, or stent.

Not everyone will experience all long-term complications or symptoms.

CoA / IAA:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 2 years

ECG/EKG, Echocardiogram:
every 2 years

Exercise Test
every 2 years

Cardiac CT/MRI:
every 3 - 5 years

Brain MRI:
An MRI of your brain is recommended **at least once** to check for a brain aneurysm. Repeat scans may be needed.

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you have CoA or IAA and are planning a pregnancy, you may require a procedure to repair it. Talk to your ACHD doctor early in your planning process.

WHAT ARE CORONARY ARTERY ANOMALIES?

Structural Issues

Coronary arteries, or coronaries, are small blood vessels that arise from the aorta and supply oxygenated blood to the heart muscle. There are two major coronaries (right and left).

Some people may have arteries with **unusual origins, courses, or connections**. These are called coronary artery anomalies.

Common types include:

- **Anomalous origin:** Coronaries arise from the wrong place in the aorta or may have an unusually shaped opening.
- **Anomalous origin from pulmonary artery** (**ALCAPA**): Coronary artery arises from the pulmonary artery instead of the aorta.
- **Coronary fistula:** An abnormal tunnel forms between the coronary artery and another heart chamber (right ventricle) or blood vessel (pulmonary artery).

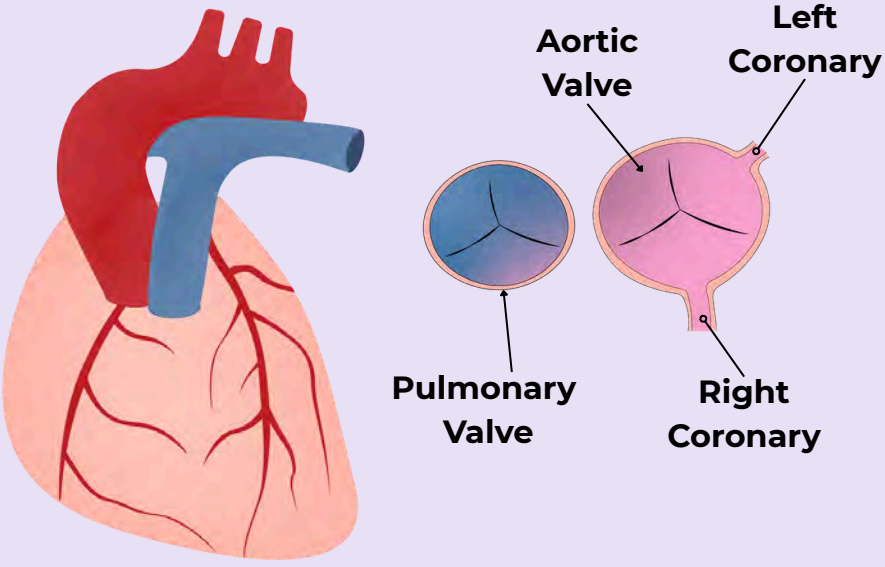
WHAT ARE CORONARY ARTERY ANOMALIES?

What Happens?

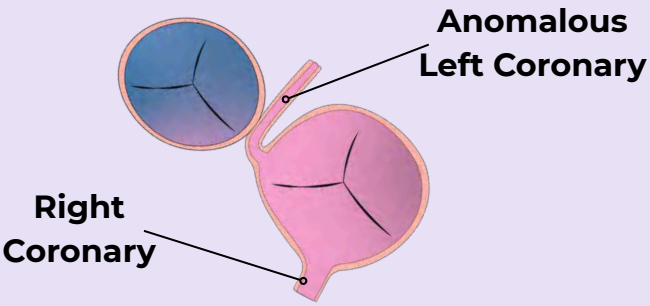
Many coronary artery anomalies do not require any treatment and are **often found by chance**.

Some types, such as a left coronary artery arising from the right side of the aorta or arising from the pulmonary artery, can be more serious.

NORMAL CORONARY ARTERIES



EXAMPLE OF A CORONARY ARTERY ANOMALY



CORONARY ARTERY ANOMALIES: WHAT TO KNOW

Statistics

Coronary artery anomalies are rare, occurring in less than **1%** of the population.

Associated With:

Structural Issues

May be experienced along with other congenital heart defects, such as:

- Transposition of the Great Arteries
- Tetralogy of Fallot
- Pulmonary atresia

It is very common for coronary artery anomalies to be discovered for the first time during teenage years or even much later in life.

CORONARY ARTERY ANOMALIES: MANAGEMENT

Not all coronary anomalies require treatment or surgery. **Treatment depends on**:

- The type of anomaly
- Whether symptoms or signs of reduced blood flow are present
- The risk of future problems

Your doctor may perform **tests**, such as EKG/ECG, echocardiogram, exercise stress test, cardiac CT, or catheterization, to assess.

Catheter Procedures

To **map the path** of the coronary arteries and check the blood flow to the heart. In some conditions, such as a coronary fistula, a **device may be placed** to close it using a catheter procedure.

Surgical Procedures

Surgery may be used to **reimplant** the artery to the correct location in the aorta. Surgery may also **bypass** an abnormally positioned artery by creating a new route for blood flow using a tissue graft. In ALCAPA (coronary arises anomalously from the pulmonary artery), a **tunnel** (baffle) is created inside the pulmonary artery and the coronary directed to the aorta.

CORONARY ARTERY ANOMALIES: LONG-TERM

Pump Issues: If the heart muscle does not receive enough blood, it may affect the ventricle's pumping function.

Valve Issues: Decreased pumping function of the ventricles can lead to enlargement of the heart chambers, which can then lead to leaking heart valves (example: mitral regurgitation).

Electrical Issues: Scar tissue in the heart or a decrease in blood supply to the heart muscle may increase the risk of abnormal fast heart rhythm (arrhythmia).

Plumbing Issues: Even after repair, some people may still experience changes in blood flow or narrowing of the coronary arteries.

Not everyone will experience all long-term complications or symptoms.

CORONARY ARTERY ANOMALIES: COMMON SYMPTOMS

Possible symptoms

Chest pain, shortness of breath, or fainting: Especially with exercise

Tiredness: Feeling more tired than usual.

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Why do these things happen?

Decreased blood supply to the heart muscle, causing decreased pumping function.

Enlarged heart chambers or **scarring** in the heart muscle.

Arrhythmias can cause palpitations.

What are the potential risks?

There may be a risk of heart attack or sudden cardiac death with certain coronary anomalies; sudden onset of chest pain and shortness of breath are common symptoms.

Not everyone will experience all long-term complications or symptoms.

CORONARY ARTERY ANOMALIES: FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG:
every year

Echocardiogram:
Every 2 years

Exercise Test, Heart Monitor:
as needed

Cardiac CT/MRI:
At the time of diagnosis; repeat as needed based on symptoms.

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs, provider preferences, and whether the anomaly is repaired or not.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS DOUBLE OUTLET RIGHT VENTRICLE (DORV)?

There are **four main types** of DORV based on the position of the main arteries (aorta and pulmonary artery) relative to the VSD.

- **DORV with subaortic VSD** (the hole is just below the aorta). This can be associated with narrowing of the pulmonary valve (pulmonary stenosis).
- **DORV with subpulmonic VSD** (the hole is just below the pulmonary artery).
- **DORV with remote VSD** (the hole is far away from both arteries).
- **DORV with doubly committed VSD** (the hole is below both arteries).

Statistics

Accounts for **~1%** of all CHD.

DORV:
COMMON SYMPTOMS

Symptoms range from decreased oxygen levels (cyanosis) to signs of heart failure (swelling or fatigue).

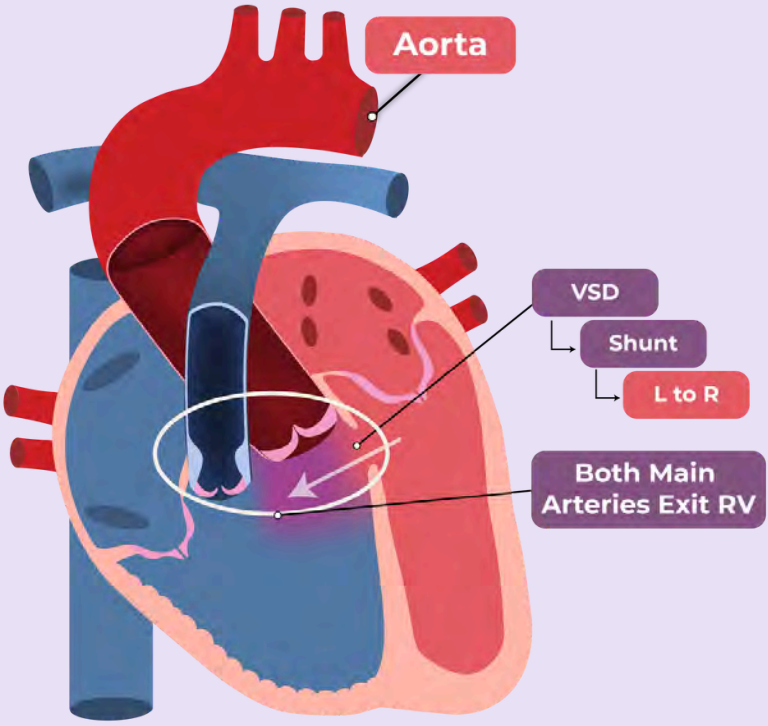
Your specific symptoms depend on:

Structural

VSD Location: Where the hole (VSD) sits in relation to the aorta and pulmonary arteries.

Valve

Valve Narrowing: Whether the pulmonary valve is narrowed (stenosis)**.**



DORV:
COMMON REPAIRS

Surgical Procedures

To connect the aorta to the left ventricle and the pulmonary artery to the right ventricle so blood flows correctly.

Rastelli Repair: The surgeon patches the hole (VSD) and uses a tube with an artificial valve (conduit) to connect the right ventricle to the pulmonary artery. **most common*

Arterial Switch: The surgeon switches the two main arteries to align them with the correct pumping chambers.

Fontan Circulation: If one ventricle is underdeveloped or the VSD is located in a difficult position, you may have a series of operations leading to a Fontan circulation.

Please see the Single Ventricle section to learn about long-term care for this circulation type.

You will be able to receive a PDF of all the conditions at the end.

DORV:
LONG-TERM

Potential issues can arise and depend on your anatomy and the surgery you've had.

Structural Issues: Valves or conduits may eventually become leaky (regurgitation) or too narrow (stenosis). If this happens, a catheter procedure or surgery may be needed to fix or replace them.

Electrical Issues: Scarring from prior surgeries can disrupt electrical signals, leading to irregular or rapid heartbeats (arrhythmias). These are often controlled with medication, ablation, or, in some cases, a pacemaker.

Not everyone will experience all long-term complications or symptoms.

DORV:
LONG-TERM

***Pump Issues*:** Over time, the lower chambers (ventricles) may pump less effectively. This can be caused by valve issues, irregular heart rhythms, or a weakened heart muscle.

EMPOWERMENT

You might hear your doctor refer to this sort of pump issue as "low ejection fraction" or "abnormal heart function".

***Plumbing Issues*:** Coronary artery disease can develop, where there is narrowing of the arteries that supply blood to the heart muscle. This is a specific risk to monitor if you had an Arterial Switch operation.

The arteries carrying **blood to the lungs** (pulmonary artery) can sometimes become **too narrow (**stenosis). If this happens, a catheter procedure may be needed to open them.

Not everyone will experience all long-term complications or symptoms.

DORV:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every year

Cardiac CT/MRI:
every 3 - 5 years

Exercise test, Heart Monitor:
as needed

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS EISENMENGER SYNDROME?

Eisenmenger syndrome is a condition that typically **develops secondary** to a congenital heart defect.

Cause: It occurs when a heart defect allows "blue" (less oxygenated) and "red" (oxygenated) blood to mix.

Timeline: When this type of defect is not repaired in childhood to separate the blood, the lungs are exposed to excess blood flow for many years.

Result: Over time, this extra blood flow often leads to Eisenmenger Syndrome.

Statistics

Occurs in **1 - 6%** of those with CHD.

Associated With:

- Endocardial Cushion/Atrioventricular septal defect (AVSD)
- Atrioventricular Septal Defect (ASD)
- Ventricular Septal Defect (VSD)
- Patent Ductus Arteriosus (PDA)
- Single Ventricle

EISENMENGER SYNDROME: WHAT TO KNOW

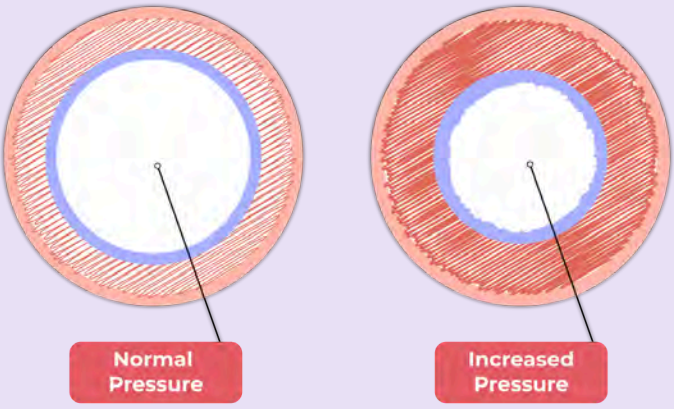
Cyanosis (Low Oxygen)

Less oxygenated blood mixes with oxygenated blood, so less oxygen reaches your body.

You may notice a bluish tint to your lips, fingers, or skin. Your oxygen levels are likely lower than normal (often below 90%) both at rest and during exercise.

Pulmonary Hypertension (High Lung Pressure)

The extra blood flow to the lungs may cause the blood vessels to become stiff and narrow.



This narrowing acts like a clogged pipe, causing pressure to build in the lung arteries and making the right side of your heart work harder.

Not all pulmonary hypertension is caused by a CHD. It could be caused by lung disease, blood clots, other heart diseases, etc.

EISENMENGER SYNDROME: LONG-TERM

Effects of Low Oxygen

- ***Cyanosis***: You might get tired or turn blue more easily than your peers.
- ***Fingers or toes could become swollen, and nails look rounder*** or more curved than normal (clubbing) – this is a way for the body to adapt to chronic low oxygen levels.
- ***Increased hemoglobin*** (secondary erythrocytosis): Hemoglobin carries oxygen in your blood. To compensate for your lower oxygen levels, your body produces extra hemoglobin.
- ***Increased risk for blood clots:*** the increased hemoglobin could sometimes make the blood "sticky", especially when you are dehydrated. This can increase the risk of clots and brain infections.
- ***Infection Risk*:** The unrepaired heart defect creates a pathway that allows bacteria to reach the brain more easily.

Not everyone will experience all long-term complications or symptoms.

EISENMENGER SYNDROME: LONG-TERM

Secondary to pulmonary hypertension you may have:

Pump Issues: Because the right side of your heart (right ventricle) pumps against high pressure in the lungs, the muscle becomes thicker and larger over time, which may reduce its pumping function.

Valve Issues: The high pressure in your lungs can put strain on the heart valves, especially on the right side (tricuspid or pulmonary valve). These valves might leak (regurgitation), leading to an enlarged heart.

Electrical Issues: Enlargement and strain on the right heart structures can lead to problems with the heart's electrical system, causing irregular heartbeats (arrhythmias).

Not everyone will experience all long-term complications or symptoms.

EISENMENGER SYNDROME: TREATMENT

Often, a surgical repair is not an option.

Because the blood vessels in your lungs are stiff and narrow, pressure builds up on the right side of your heart. The hole actually acts as a "safety valve," allowing blood to escape to the left side to relieve this pressure.

Why wouldn't you close the hole?
In most cases, if surgeons were to close this hole, the pressure would have nowhere to go. This would cause the right side of the heart to fail.

Management: Since surgery cannot usually safely fix the defect, the focus of your care shifts to medical management. We will cover these aspects next.

In some very specific cases, a lung transplant or a heart-lung transplant might be considered.

EISENMENGER SYNDROME: MANAGEMENT

Manage Iron Levels

- Iron is the building block for hemoglobin, and levels should be tightly controlled
- Your doctor will monitor your iron stores (ferritin levels) and may prescribe supplements for short intervals.
- You need enough iron to carry oxygen, but avoiding excess is crucial to prevent your blood from becoming too "thick."

Oxygen Support

Supplemental oxygen (mask or cannula) can help reduce strain and improve your quality of life, but likely won't bring your oxygen levels back to "normal".

EMPOWERMENT

Ask your ACHD cardiologist if home oxygen would benefit you.

EISENMENGER SYNDROME: MANAGEMENT

Reduce Blood Clot Risk

- Stay well hydrated.
- If you plan to travel to high-altitude areas, take extra precautions, such as drinking plenty of water and carrying oxygen as needed.

Medication Management

Sildenafil, macitentan, bosentan, or other pulmonary vasodilators can help relax the blood vessels in your lungs, making it easier for blood to flow through them.

These medications are usually helpful for improving exercise tolerance or oxygen levels, even though your lung pressure may not get to a normal level.

Beta-blockers, SGLT2, or other medications may be recommended to improve your heart function and quality of life.

Blood thinners (aspirin, warfarin) may be recommended to reduce blood clot risk.

EISENMENGER SYNDROME: MANAGEMENT

Phlebotomy (Blood Removal)

Routine phlebotomy is not generally recommended. **In rare cases**, doctors remove some of your blood to lower hemoglobin levels.

CAUTION: *If this is suggested,* ***ALWAYS consult your ACHD doctor*** *first to ensure it is actually beneficial for you.*

IV Safety (Air Filters)

Air bubbles in an IV line can cross the hole in your heart and travel to the brain (stroke risk).

EMPOWERMENT

Whenever you get an IV, **ask for an air filter** on the line. Filters act like a tiny screen within the IV line, preventing unwanted air bubbles or particles from crossing the hole in your heart and reaching your brain.

EISENMENGER SYNDROME: FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 6 - 12 months

ECG/EKG, Echocardiogram:
every year

Exercise Test:
every 6 - 12 months

Labs:
(iron levels, ferritin & hemoglobin)
every year

Heart Monitor, Cardiac CT/MRI, Cath:
as needed

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

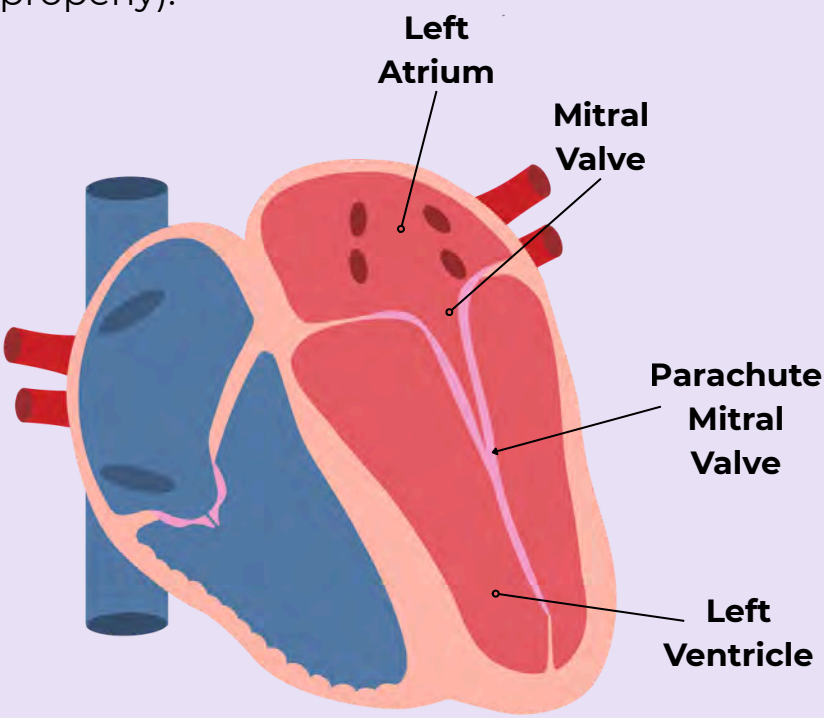
WHAT ARE MITRAL VALVE CONDITIONS?

Mitral valve conditions affect the valve located between the upper and lower chambers on the left side of the heart (the left atrium and left ventricle).

Structural Issues

Mitral valve stenosis (Narrowing): Conditions that cause mitral stenosis include parachute mitral valve, mitral arcade, narrow mitral valve, and supra-mitral ring.

Mitral valve regurgitation (Leaking): The valve does not close tightly. Conditions include mitral valve prolapse (floppy valve) or mitral cleft (the valve did not develop properly).



WHAT ARE MITRAL VALVE CONDITIONS?

What Happens?

The mitral valve controls blood flow from the left atrium into the left ventricle, which is the heart's main pumping chamber.

Mitral Valve Stenosis (Narrowing): If the mitral valve is narrowed, blood cannot flow easily from the left atrium into the left ventricle. When severe, this can cause blood to back up into the left atrium and lungs, increasing pressure in the lungs and making it harder to breathe. The heart must work harder to pump blood through the valve.

Mitral Valve Regurgitation (Leaking): If the mitral valve is leaky, blood flows backward from the left ventricle into the left atrium instead of moving forward to the body. This means the heart has to pump extra blood with each beat to make up for what leaked backward, forcing the heart to work harder.

If you have mitral atresia, you likely have single ventricle anatomy. Please refer to the Single Ventricle section.

If you have Shone's Complex, please refer to the Shone's Complex section.

You will be able to receive a PDF of all the conditions at the end.

MITRAL VALVE:
WHAT TO KNOW

Associated Conditions:

May occur alongside other **left-sided heart conditions:**

1. Shone's Complex
2. Coarctation of Aorta: narrowing in the main artery leaving the heart
3. Subaortic stenosis: narrowing below the aortic valve
4. Parachute mitral valve: the valve's support structures (chordae) attach to only one muscle instead of being spread across two
5. Supravalvular mitral ring - extra tissue above the valve that causes narrowing

Statistics

Mitral valve conditions alone (that are not associated with other CHD) are rare, each making up **less than 1%** of all CHD.

Many people with mitral valve conditions have no symptoms, and the condition may be discovered incidentally during a routine exam or imaging test for another reason.

MITRAL VALVE: PROCEDURES

If the narrowing (stenosis) is severe enough to restrict blood flow to the left ventricle, or if the leak (regurgitation) is severe enough to strain the heart, you may need a procedure to repair or replace the mitral valve. Repair is always preferred when possible, as it preserves your natural valve.

Surgical Procedures

Repair: The surgeon fixes the existing valve, which may include tightening the ring-like structure around the valve (annulus), reconstructing the leaflets, or repairing or replacing the support structures (chordae or papillary muscles).

Replacement: If the valve is too abnormal or damaged to repair, it is replaced with a new artificial valve.

Catheter Procedures

Transcatheter mitral valve repair (TMVr) is used to repair a leaky valve. A clip is placed through a catheter to bring the valve leaflets closer together, reducing leakage.

Transcatheter mitral valve replacement (TMVR) is used to treat both leaky valves (regurgitation) and narrow valves (stenosis). A new valve is placed inside the old one using a catheter, without open-heart surgery.

MITRAL VALVE:
LONG-TERM

Not everyone will experience these long-term complications, but it's important to be aware of potential issues that may develop over time.

Valve Issues: Narrowing (stenosis) can recur after repair, or the valve may develop a leak (regurgitation). Artificial valves may wear out or malfunction over time and may need repair or replacement

Structural issues: If the mitral valve is narrowed or leaky, blood can back up into the left atrium and lungs, causing increased pressure in the lungs (pulmonary hypertension).

Pump Issues: With mitral stenosis (narrowing), the increased lung pressure can strain the right ventricle, weakening its pumping function over time. With mitral regurgitation (leaking), the left ventricle becomes enlarged from pumping extra blood with each beat, which can weaken its pumping function over time.

Electrical Issues: An enlarged left atrium increases the risk of arrhythmias, such as atrial flutter or fibrillation. These may require medication, ablation, or blood thinners (anticoagulation).

MITRAL VALVE:
COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Shortness of breath: Particularly with exercise or laying flat.

Swelling: Extra fluid in your legs, belly, or around the eyes.

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Why do these things happen?

Tiredness, shortness of breath: When the mitral valve is narrowed or leaky, blood backs up into the lungs, causing pressure and fluid buildup. The blood vessels in the lungs may become stiff, making it harder for blood to carry enough oxygen to your body. This can leave you feeling tired and short of breath, even with light activity.

Swelling: When pressure in the lungs increases, the right ventricle has to work harder and may become weak with time. This may cause fluid to back up into the body and lead to selling.

Irregular heartbeats: An enlarged left atrium from the valve problem can disrupt the heart's electrical system, causing it to beat irregularly or too fast.

MITRAL VALVE CONDITIONS: FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 1 - 2 years

ECG/EKG
every 1 - 2 years

Echocardiogram:
every 2 years

Exercise Test:
every 2 years

Cardiac MRI/CT:
if needed for further evaluation or planning intervention

**Based on the 2018 or 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

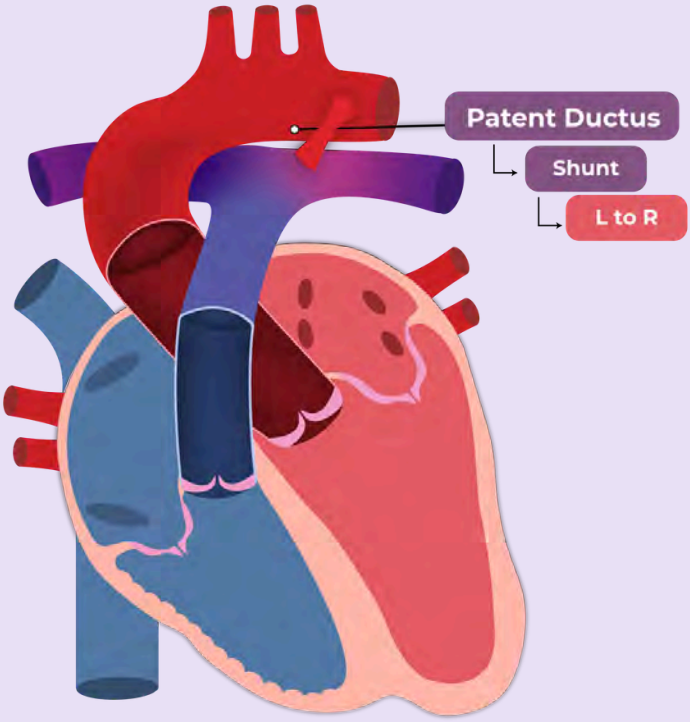
WHAT IS A PATENT DUCTUS ARTERIOSUS (PDA)?

Structural Issues

The ductus arteriosus is an important blood vessel that **supports normal circulation** in the fetus before birth. It usually closes on its own within a few days after birth. When it **stays open** (patent), it is known as a patent ductus arteriosus.

What happens?

PDA causes some blood to flow from the aorta into the pulmonary artery — **left (L) to right (R) shunt**.



PDA:
WHAT TO KNOW

Statistics

PDA accounts for **5 - 10 %** of CHD cases.

It is not uncommon for it to be detected for the first time in adulthood.

Associated With:

- Severe Pulmonary Valve Stenosis
- Coarctation of Aorta (CoA)
- Transposition of the Great Arteries
- Hypoplastic Left Heart Syndrome (HLHS)
- Interrupted Aortic Arch (IAA)
- Pulmonary Atresia

If you have a PDA along with other CHD defects, please review the sections for those conditions as well.

You will be able to receive a PDF of all the conditions at the end.

PDA:
MANAGEMENT

Most PDAs **close on their own** in childhood.

If your PDA **doesn't close** by adulthood and your doctor determines it should be **repaired**, there are two possible treatment options: catheter procedure or surgery.

Catheter Procedure

A less invasive procedure where a device (such as a duct occluder or coil) is used to close the hole.

Surgical Procedure

If catheter closure is not possible because of the PDA's size or shape, a surgical procedure may be done to tie it off.

**Catheter procedure is more common than surgery*

Did You Know?

In some CHD conditions, keeping the ductus open after birth can be lifesaving because it helps maintain blood flow until the heart can be repaired.

SMALL UNREPAIRED **PDA:** COMMON ISSUES & SYPMPTOMS

Long-Term

Structural Issues: Because blood flows through the small PDA at high velocity, it can irritate and damage the inner lining of the vessel. This can create a spot where bacteria can attach and cause an infection (endocarditis).

Possible Symptoms

Your doctors may hear a loud **heart murmur** (an extra “whoosh” or “swish” sound when they listen to your heartbeat).

Even if you have no symptoms, your doctor may recommend repair to remove the lifelong risk of endocarditis.

Not everyone will experience all long-term complications or symptoms.

SMALL UNREPAIRED **PDA:** FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 1 - 5 years

ECG/EKG, Echocardiogram:
every 1 - 5 years

Heart Monitor, Exercise Test:
as needed

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

LARGE UNREPAIRED PDA: LONG-TERM

Structural Issues: Over time, extra blood flows to the lungs and back to the left side of the heart. This leads to enlargement of the left heart chambers.

Pump Issues: The left side of the heart has to handle more blood, so it has to pump harder. This can cause the heart chambers (atria or ventricles) to enlarge and weaken.

Electrical Issues: Enlarged upper chambers (atria) can lead to abnormal heart rhythms, such as atrial flutter and atrial fibrillation.

Plumbing Issues: Excess blood flow to the lungs increases pressure in the pulmonary arteries (pulmonary hypertension). This leads to stiffening of the arteries, which can make the right side of the heart (right ventricle) pump harder into the lungs.

Valve Issues: As the extra blood from the lungs returns to the left side of the heart, the left ventricle enlarges. This can stretch the mitral valve, causing it to leak.

Infection Risk: There could be a risk of infection in the heart valves and heart lining.

Not everyone will experience all long-term complications or symptoms.

LARGE UNREPAIRED **PDA:** COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Why do these things happen?

Tiredness: If blood pressure in your lungs becomes too high, the blood vessels may become stiff, making it hard for the blood to carry enough oxygen to the body. Your oxygen levels might decrease, and you might feel tired easily even with small activity.

Swelling: Can occur due to weak heart function, leaky valves, or high lung pressure.

You may not have any symptoms, and your PDA might be found by chance.

LARGE UNREPAIRED PDA: TREATMENT PATHS

If Lung Pressures are Normal

- The primary treatment is **repairing the blood vessel** (usually done by a catheter procedure)
- You might need **medications** to get **rid of the extra fluid** or help your heart pump more effectively
- **Rhythm problems** can be treated with medications. An ablation may be performed in rare cases.

If Lung Pressures are High

- You may need **medications** to help **lower lung pressures**, such as sildenafil or macitentan.
- PDA **repair is often delayed** until the pressure in your lungs improves.
- You may need **blood thinners** to lower your risk of stroke.
- You might also need **medications** that help your **heart pump** more effectively

LARGE UNREPAIRED **PDA:** FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 6 - 12 months

ECG/EKG, Echocardiogram:
every year

Heart Cath:
(To assess lung pressures & response to medications)
every 6 - 12 months

Exercise Test:
every 1 - 2 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

REPAIRED PDA:
FOLLOW-UP

If you did NOT need treatment for high lung pressure before repair, this page applies to you.

Most people do well after the PDA is repaired, assuming you had no other heart-related problems.

Recommendations*

Routine Cardiologist Visits:

(outpatient)
every ~5 years

Primary Care Visits:

You can continue to follow up with your primary care doctor. Make sure your doctors know about your heart history.

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

REPAIRED PDA:
FOLLOW-UP

If you received treatment for HIGH lung pressures before repair, this page applies to you.

After repair, you may need to continue taking some lung pressure medications (sildenafil, macitentan), blood thinners, or heart rhythm medicines.

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 1 - 2 years

ECG/EKG, Echocardiogram:
every 1 - 2 years

Heart Monitor, Cath, Exercise Test:
as needed

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

PULMONARY VALVE CONDITIONS

In this section, we will review two common pulmonary valve conditions.

First, we will review **Pulmonary Atresia**. Then, we will review **Pulmonary Stenosis**.

If you want to skip to the Pulmonary Stenosis section, click through to **page 99**.

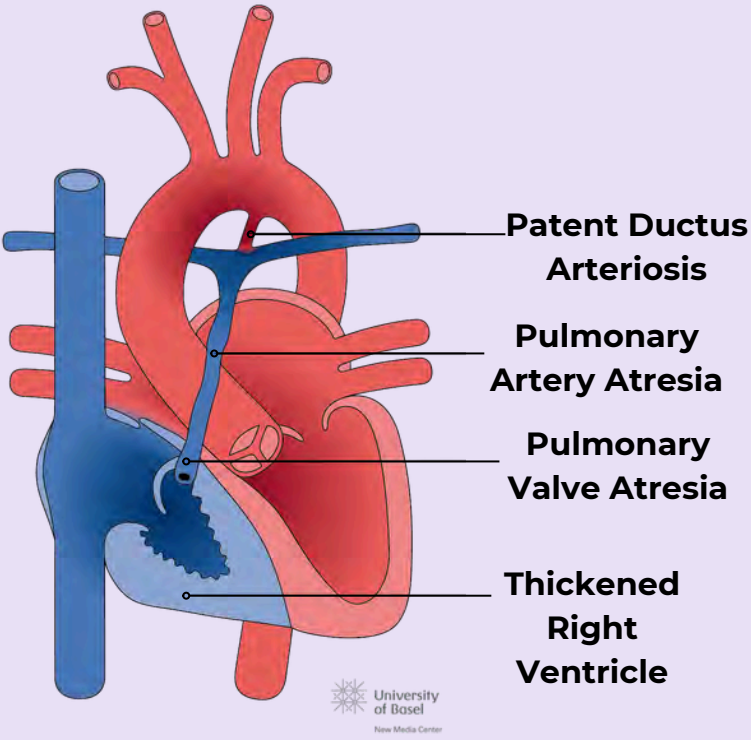
WHAT IS PULMONARY ATRESIA?

Structural Issues

The valve and/or artery on the right side of the heart (the pulmonary valve and artery) are not well-formed and so are either small or almost absent.

Pump Issues

Since the pulmonary valve/artery cannot function, it limits the right ventricle's ability to pump blood efficiently into the lungs.



Statistics

Pulmonary atresia is rare, making up **less than 1%** of all CHD.

PULMONARY ATRESIA: WHAT TO KNOW

Pulmonary Atresia can occur **with or without a hole** in the heart:

- Pulmonary atresia **with intact ventricular septum** (PA/IVS) - no hole present
- Pulmonary atresia **with ventricular septal defect** (PA-VSD) - hole is present

If you have PA-VSD, your outcomes may be similar to those with Tetralogy of Fallot (See Tetralogy of Fallot section).

If you have pulmonary atresia with a single ventricle or have had Fontan surgery, your outcomes may be similar to those with a single ventricle (See Single Ventricle section).

*The rest of **this section focuses on PA/IVS** (no hole) in patients who did not have single ventricle anatomy or Fontan-related issues.*

You will be able to receive a PDF of all the conditions at the end.

PULMONARY ATRESIA: EARLY PROCEDURES TO IMPROVE BLOOD FLOW

You may have undergone one or more **procedures** in childhood to help blood reach your lungs. These procedures improve blood flow and oxygen levels but do not fully correct the heart defect. They are often used as **temporary** solutions in childhood when a complete repair is not yet possible.

Types of Procedures You May Have Had:

Catheter Procedures

Stent placement: A small mesh tube (stent) is placed in the natural blood vessel that connects the aorta and pulmonary artery (called the ductus arteriosus) to keep it open and allow blood flow to lungs.

Balloon Valvuloplasty: If feasible, doctors may use a **balloon** procedure to stretch open the pulmonary valve.

Surgical Procedure

A tube (called as **shunt)** is placed to create a connection between the aorta (blood vessel leaving the heart) and the pulmonary artery (blood vessel going to lungs). This allows blood to bypass the narrowed valve and flow to the lungs. Examples: BT shunt or central shunt.

PULMONARY ATRESIA: MORE COMPLETE REPAIR PROCEDURES

More complete procedures often follow the early blood flow procedures described previously. These are more **permanent repairs** to the heart and are the final step(s) for complex conditions that require staged surgeries.

Surgical Procedure

Patch reconstruction: The narrowing of the pulmonary valve is opened up and repaired by placing a patch near the valve to widen it for better blood flow.

RV-PA Conduit placement: A tube containing a valve (called RV-PA conduit) is placed to connect the right ventricle (RV) and the pulmonary artery (PA). This creates a new more effective pathway for blood to flow to the lungs.

Re-intervention:

Overtime, the conduit can narrow, or the patch/valve can wear out. In such cases, you may need either *a surgical procedure to replace the conduit or valve, or a catheter procedure to place a new valve inside the existing conduit.*

PULMONARY ATRESIA: LONG-TERM

Not everyone will experience these long-term complications, but it is important to be aware of potential issues that may develop over time.

***Valve Issues*:** Conduits and replacement valves do not grow with your body and can eventually wear out. A new pulmonary valve may need to be placed if there is significant leaking or narrowing.

Structural Issues: The right ventricle can become thick (from narrowed pulmonary valve or artery) or weak (from leaky valve).

Pump Issues: The right ventricle's ability to pump effectively may decrease over time, especially if there is narrowing or leaking in the conduit or valve.

Plumbing Issues: High pressure in the right ventricle from a narrowed valve can result in the development of abnormal connections between the coronary arteries (which supply blood to the heart muscle) and the heart chambers, called coronary cameral fistulas (CCF). These connections can affect how well the heart muscle receives oxygen. If needed, these may be treated with a catheter procedure.

PULMONARY ATRESIA: LONG-TERM

Not everyone will experience these long-term complications, but it is important to be aware of potential issues that may develop over time.

***Electrical Issues*:** Scar tissue from surgery and extra strain on the heart may lead to irregular heartbeats (arrhythmias), such as atrial fibrillation or ventricular tachycardia. This may require treatment with medications, a procedure to reset the heart rhythm (cardioversion), or a procedure to eliminate the source of the irregular rhythm (ablation).

Infection Risk: There is an increased risk of infection (endocarditis) if you have a conduit or artificial valve. This happens because bacteria can attach to these materials more easily than natural tissue. Preventive antibiotics before certain dental or medical procedures may be recommended.

PULMONARY ATRESIA
COMMON SYMPTOMS

Possible Symptoms

Tiredness, shortness of breath: Feeling more tired than usual, especially with activity or exercise.

Swelling: Extra fluid in your legs, belly, or around the eyes.

Irregular heartbeats: Feeling your heart racing or extra beats (palpitations).

Why do these things happen?

Tiredness, shortness of breath: Narrowing of the pulmonary valve or conduit can cause extra strain on the right ventricle. This can weaken its pumping ability over time.

Swelling: When the right ventricle becomes strained or weakened, pressure can build, causing fluid to back up into the body.

Irregular heartbeats: Scar tissue from previous surgeries and ongoing strain on the heart can disrupt the heart's electrical system, leading to irregular or rapid heartbeats.

Not everyone will experience all long-term complications or symptoms.

PULMONARY ATRESIA:
FOLLOW UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every year

Exercise Test, Heart Monitor:
As needed

Cardiac CT/MRI:
every 3 - 5 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS PULMONARY STENOSIS?

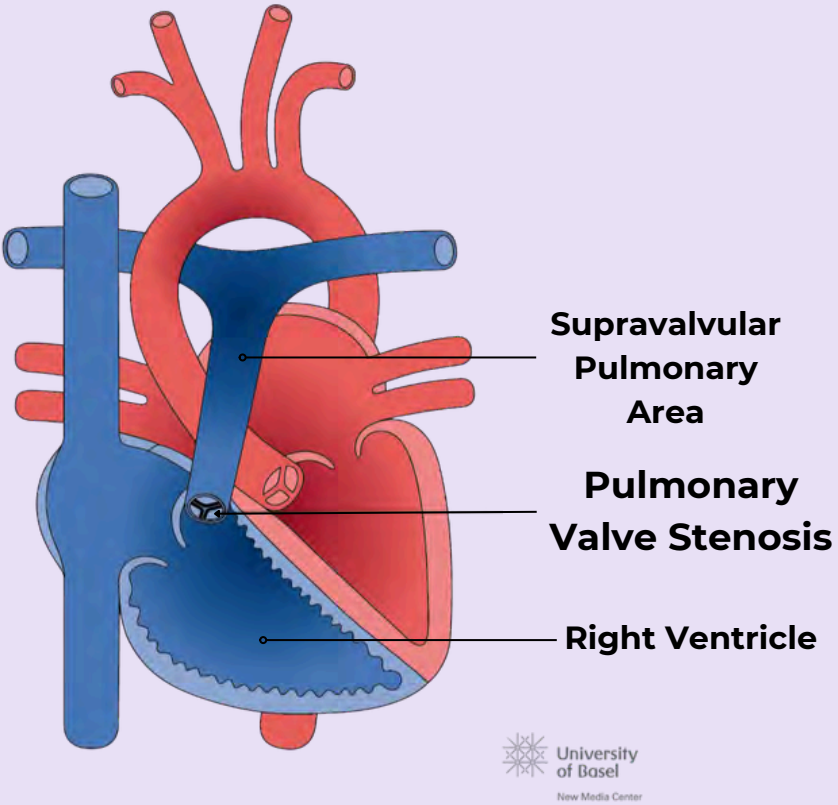
Structural Issues

The pulmonary valve or the area around it is narrowed, making it harder for blood to flow from the heart into the lungs. When this affects the valve, it is called **pulmonary valve stenosis**

If it affects the blood vessel just above the pulmonary valve, it is called **supravalvular pulmonary stenosis.**

Pump Issues

This forces the right side of the heart to work much harder to push blood through the tight area.



PULMONARY STENOSIS: WHAT TO KNOW

This condition may occur in isolation or be associated with other heart defects.

Associated With:

Structural Issues

Atrial Septal Defect (ASD): A hole between the upper chambers (atria).

Ventricular Septal Defect (VSD): A hole between the lower chambers (ventricles).

Genetics

Williams Syndrome, Noonan Syndrome, or **Alagille Syndrome**: may be associated with narrowing above the valve (supra-valvar stenosis).

Statistics

Pulmonary stenosis makes up about **8 - 10%** of all congenital heart disease.

PULMONARY STENOSIS: MANAGEMENT

Pulmonary Stenosis is often a mild condition. In many cases, it may not cause symptoms and can be monitored without treatment. Sometimes, even severe narrowing may not be detected until later in childhood or adulthood.

When Treatment Is Needed

If the narrowing is severe and causes symptoms, it typically presents in early childhood and often requires intervention to relieve the obstruction.

Treatment Options:

Catheter Procedure

A small balloon is threaded through a blood vessel to the narrowed valve and inflated to stretch it open **(balloon valvuloplasty)**. This is the most common first treatment.

Surgical Procedures

Surgival valvotomy / Brock procedure may be performed to cut open and relieve the narrowing. This is typically used when a catheter procedure is not possible or has not been successful.

PULMONARY STENOSIS: MANAGEMENT

What Happens After The Initial Treatment?

After relieving the narrowing in the pulmonary valve (by catheter or surgical procedure), the pulmonary valve may develop a leak (pulmonary regurgitation). This leaking may be mild at first but can worsen over time, causing the right ventricle to become enlarged.

Pulmonary Valve Replacement

If the leaking becomes severe and the right ventricle enlarges significantly, valve replacement becomes necessary. This can typically be done with a catheter-based procedure, but valve replacement may be performed surgically, depending on the anatomy.

PULMONARY STENOSIS: LONG-TERM

Not everyone will experience these long-term complications, but it's important to be aware of potential issues that may develop over time

Valve Issues: The pulmonary valve may develop leakage (regurgitation) over time after a catheter or surgical procedure, or it may become more narrowed (stenosis) if not previously treated. This can increase the workload on the right side of the heart.

***Pump Issues*:** With time, the right ventricle can become enlarged from a leaky valve or thickened from pushing against a narrowed valve. These changes can weaken the heart's pumping function.

Electrical Issues: Changes in heart structure and long-term strain can lead to irregular heartbeats (arrhythmias), including atrial flutter, atrial fibrillation, or ventricular tachycardia.

Infection Risk: There is a risk of bacterial infection of the heart valve (endocarditis), especially after valve procedures.

PULMONARY STENOSIS:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 2 years

ECG/EKG, Echocardiogram:
every 2 years

Exercise Test:
every 2 years

Heart Monitor, Cardiac CT/MRI:
as needed

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

Anomalous Pulmonary Venous Return:
Partial (PAPVR) & Total (TAPVR)

Anomalous pulmonary venous return could be categorized as “partial” or “total”.

Partial (PAPVR) is more common.

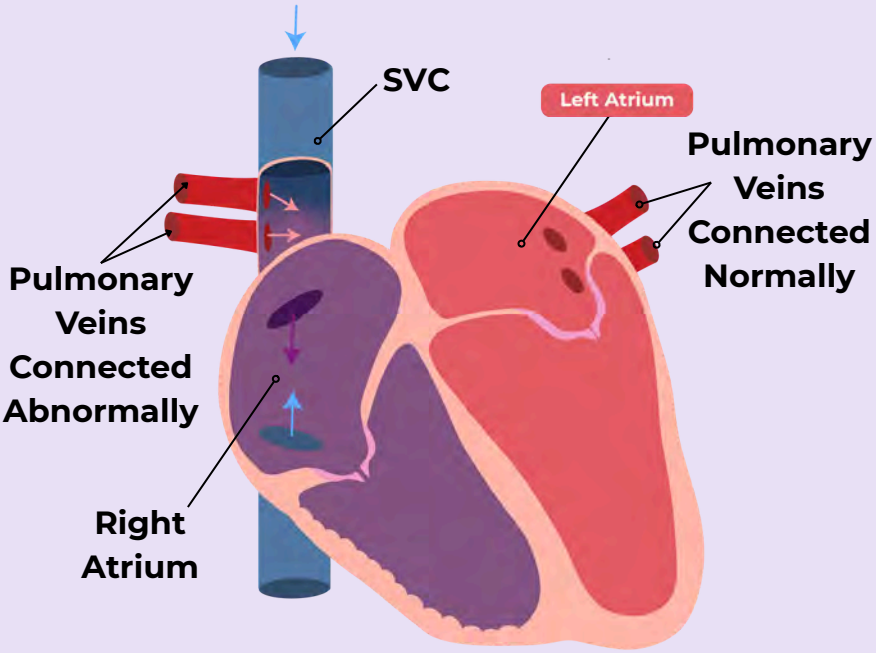
Total (TAPVR) is very rare and repaired in childhood. We will focus on long-term aspects of TAPVR.

In this section, we will first go over PAPVR and then TAPVR. If you would like to skip to TAPVR, click through to **page 113**.

WHAT IS PARTIAL ANOMALOUS PULMONARY VENOUS RETURN (PAPVR)?

Structural Issues

Some of the pulmonary veins (but not all four) carrying oxygenated blood from the lungs abnormally connect to the right side of the heart (superior vena cava (SVC), right atrium, or inferior vena cava) instead of connecting to the left atrium.



What Happens?

Because of these abnormal connections, oxygenated blood mixes with less oxygenated blood. This causes extra blood flow to the right heart and lungs, leading to enlargement of the right atrium and right ventricle. This can increase strain on the right side of the heart.

PAPVR:
WHAT TO KNOW

Statistics

PAPVR occurs in about **0.2 - 0.7%** of the general population.

80 - 90% of patients with PAPVR also have an ASD.

Associated With:

Structural Issues

Atrial Septal Defect (ASD): A hole between the upper chambers (atria).

Scimitar Syndrome: A rare type of PAPVR where a pulmonary vein from the right lung drains into the inferior vena cava (a large vein bringing blood back to the heart from the lower part of the body). This condition is associated with an underdeveloped or smaller right lung.

PAPVR: MANAGEMENT

Surgery is recommended if there is significant extra blood flow to the lungs, causing enlargement of the right heart structures and strain on the heart.

In this case, **symptoms** such as shortness of breath or decreased exercise tolerance are present.

Surgical Procedure

The lung veins are redirected to the left heart through the hole in the top chambers (Warden procedure) or may be directly connected to the left atrium.

Catheter Procedure

A covered stent is placed to redirect the pulmonary veins to the left heart.

Scimitar Syndrome *may require more complex surgical repair and ongoing monitoring due to the associated abnormalities.*

UNREPAIRED **PAPVR:** LONG-TERM

Structural Issues: The extra blood flow to the lungs can cause the pulmonary arteries to become thickened and stiff (pulmonary vascular disease).

Pump Issues: The right side of the heart becomes enlarged and overworked as it pushes against increased lung pressures. It presents with heart failure symptoms such as shortness of breath, swelling, and fatigue.

Valve Issues*:* Due to pressure buildup in the lungs and enlargement of the right ventricle, the tricuspid valve may leak (regurgitation). This may lead to strain on the right heart and worsen the valve regurgitation.

Electrical Issues: Right heart enlargement can affect the heart's electrical system, leading to irregular heart rhythms (arrhythmias). These may feel like palpitations or skipped beats.

Not everyone will experience all long-term complications or symptoms.

UNREPAIRED **PAPVR:** FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 2 years

ECG/EKG, Echocardiogram:
every 2 years

Exercise test, Cardiac CT/MRI:
every 3 - 5 years

Heart Cath:
as needed to assess pressure in lungs and if planning for an intervention

The above may be needed more often if there are signs of high lung pressure or right heart failure

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

REPAIRED PAPVR: LONG-TERM

Structural Issues: There is a risk of narrowing in the pulmonary vein at the location of the surgical repair. Symptoms can range from none at all to shortness of breath or chest discomfort.

There is a risk of narrowing of the superior vena cava after the Warden procedure. This may present as facial swelling or headaches.

Electrical Issues: There is a risk of irregular heartbeats (palpitations), such as atrial fibrillation, due to the dilated heart chambers or scarring from the surgical repair.

Not everyone will experience all long-term complications or symptoms.

REPAIRED PAPVR:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 3 - 5 years

ECG/EKG, Echocardiogram:
every 3 - 5 years

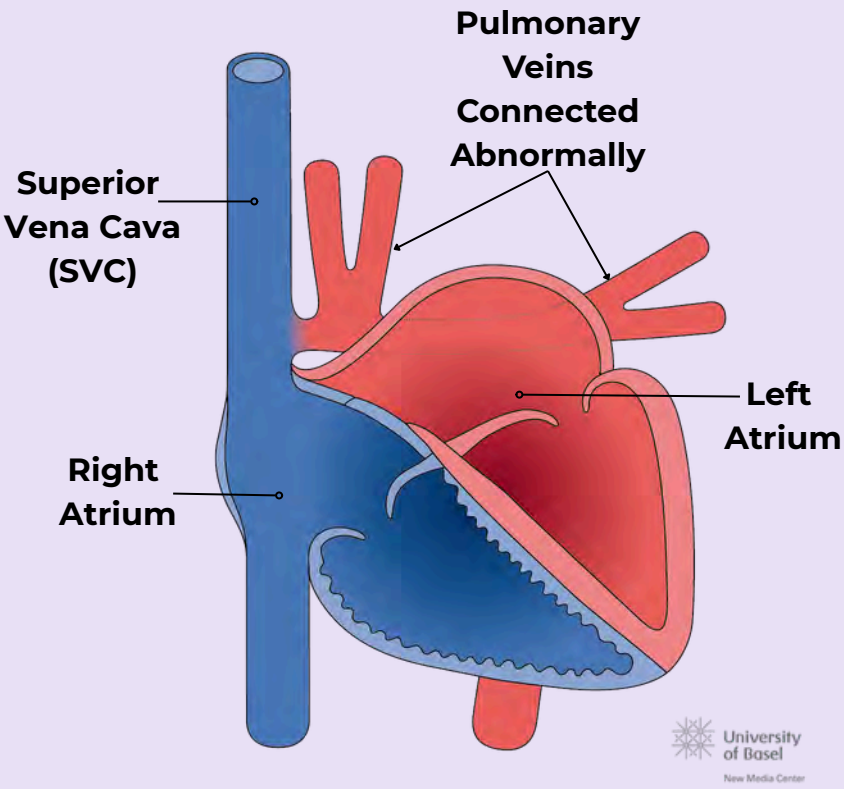
**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)?

Plumbing Issues

All four pulmonary veins carrying oxygenated blood from the lungs flow into the right side of the heart instead of the left side.



Statistics

TAPVR is rare, making up about **0.7 - 1.5%** of all CHD.

WHAT IS TOTAL ANOMALOUS PULMONARY VENOUS RETURN (TAPVR)?

There are **4 types**, depending on the location of the connection:

- **Supracardiac** TAPVR: veins connect above the heart to the superior vena cava (SVC) **most common*
- **Cardiac** TAPVR: veins connect behind the heart
- **Infracardiac** TAPVR: veins connect below the heart
- **Mixed** TAPVR: combination of the above types

Structural Issues

Due to abnormal connections of pulmonary veins to right heart structures, there is mixing of less oxygenated blood and oxygenated blood in the right heart, which leads to low oxygen levels (cyanosis). This often requires a surgical repair in early infancy.

Associated With:

Structural Issues

This condition may occur in isolation (which is very rare) or be associated with other complex heart conditions, such as heterotaxy syndrome.

TAPVR:
REPAIR & LONG-TERM IMPLICATIONS

Surgical Procedure

Surgery is usually done in the first few months of life to connect the pulmonary veins to the left atrium.

You will need close follow-up with an ACHD cardiologist to evaluate for the following long-term issues, such as:

Plumbing Issues: You may develop narrowing (stenosis) at the site where the pulmonary veins are connected to the left atrium due to scar formation. This is called pulmonary vein obstruction and may lead to pressure buildup in the lungs (pulmonary hypertension).

Electrical Issues: There is a risk of abnormal heart rhythm (atrial flutter or atrial fibrillation) due to scars at the surgical repair site.

Not everyone will experience all long-term complications or symptoms.

TAPVR:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 3 - 5 years

ECG/EKG, Echocardiogram:
every 3 - 5 years

Heart Monitor, Exercise Test, Cardiac CT/MRI:
as needed

The above may be needed more often if you have pulmonary vein stenosis or arrhythmia.

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

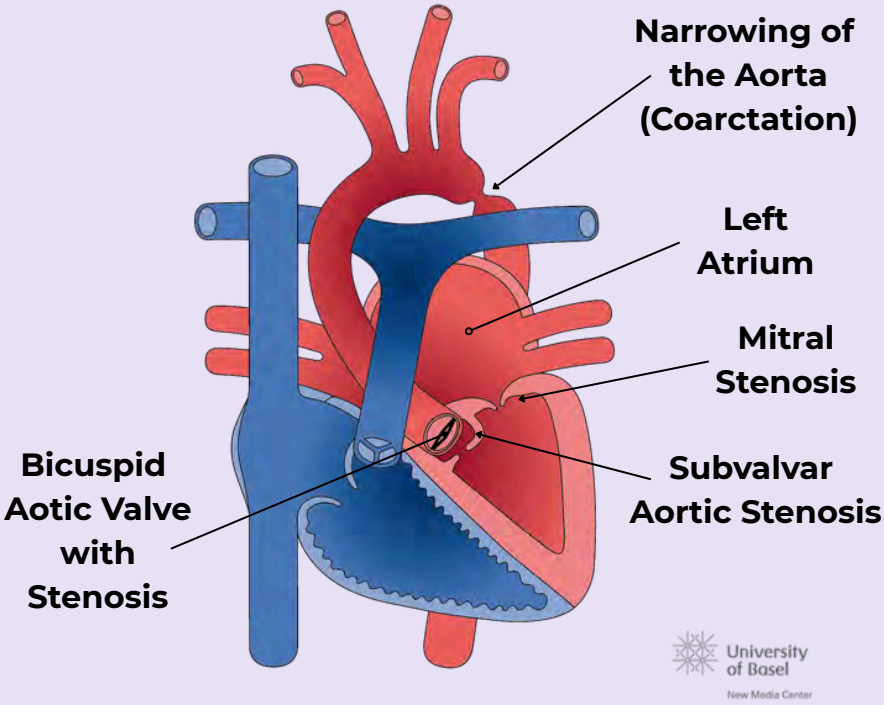
WHAT IS A SHONE'S COMPLEX?

Structural

Shone's complex is a condition where several **structures** on the **left side of the heart** are **narrowed.** Patients may have some or all of the following:

- **Coarctation of the aorta:** narrowing or constriction of the aorta, the large vessel that carries blood from the heart to the body.
- **Valvular and/or subvalvular aortic stenosis:** narrowing of the aortic valve and/or the area below the aortic valve. The aortic valve might be abnormal (bicuspid aortic valve).
- **Mitral stenosis**: a narrowed mitral valve (sometimes referred to as a parachute mitral valve), the valve between the left upper and lower chambers
- **Supravalvar mitral ring**: a thin layer of tissue is formed above the mitral valve within the left atrium. This may restrict blood flow through the mitral valve into the left ventricle.

SHONE'S COMPLEX: WHAT TO KNOW



What Happens?

Defects that involve the mitral valve interfere with the flow of blood into the left ventricle. Defects that involve the aortic valve or aorta make it harder for the left ventricle to pump blood to the body. This can lead to:

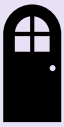
- Extra strain on the left ventricle
- The left ventricle muscle may thicken
- The left ventricle size may be smaller than normal

Statistics

Less than 1% of people with congenital heart disease.

SHONE'S COMPLEX:
LONG TERM

Structural Issues: Recurrent narrowing may occur at previous repair sites due to tissue regrowth.



Valve Issues: Mitral or aortic valve leakage (regurgitation) or narrowing (stenosis).

Pump Issues: Structural or valve issues can lead to weakening or stiffness of the left ventricle.

Electrical Issues: Surgical repairs or weakening of the ventricle can lead to arrhythmias or heart blocks.

Plumbing Issues: The aorta may become narrow (coarctation) or bulge (aneurysm) after initial repair. Narrowing of the aorta can increase the risk of high blood pressure (hypertension).

***Infection Risk*:** There is a risk of heart valve infection (endocarditis).

Not everyone will experience all long-term complications or symptoms.

SHONE'S COMPLEX: MANAGEMENT

Surgery or catheter-based interventions are often needed, depending on the extent of narrowing. These may include:

Catheter Procedure

A balloon or stent is placed to repair coarctation of the aorta or to open up the aortic valve.

Surgical Procedures

To remove the extra tissue above or below the mitral or aortic valves.

Repair or replace the mitral valve or aortic valve if it is too narrow.

Repair of the coarctation of the aorta may also be needed.

SHONE'S COMPLEX: COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid buildup in lungs or legs.

Shortness of breath / difficulty breathing

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

High blood pressure: If not well controlled, you may experience headaches or nosebleeds.

Why do these things happen?

Tiredness, Swelling, Shortness of Breath, Irregular Heartbeats: Can occur from abnormal valves, weak heart function, leaky valves, or high lung pressure that backs up fluid into the lungs, causing these symptoms.

High Blood Pressure: Narrowing of the aorta can lead to stiff blood vessels and increase the risk of high blood pressure.

Not everyone will experience all long-term complications or symptoms.

SHONE'S COMPLEX:
FOLLOW UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 1 - 2 years

ECG/EKG, Echocardiogram:
every 1 - 2 years

Exercise Test:
every 2 years

Cardiac CT/MRI:
every 3 - 5 years

Brain MRI:
If you have coarctation of aorta, a brain MRI is recommended **at least once** to check for a brain aneurysm.

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences*

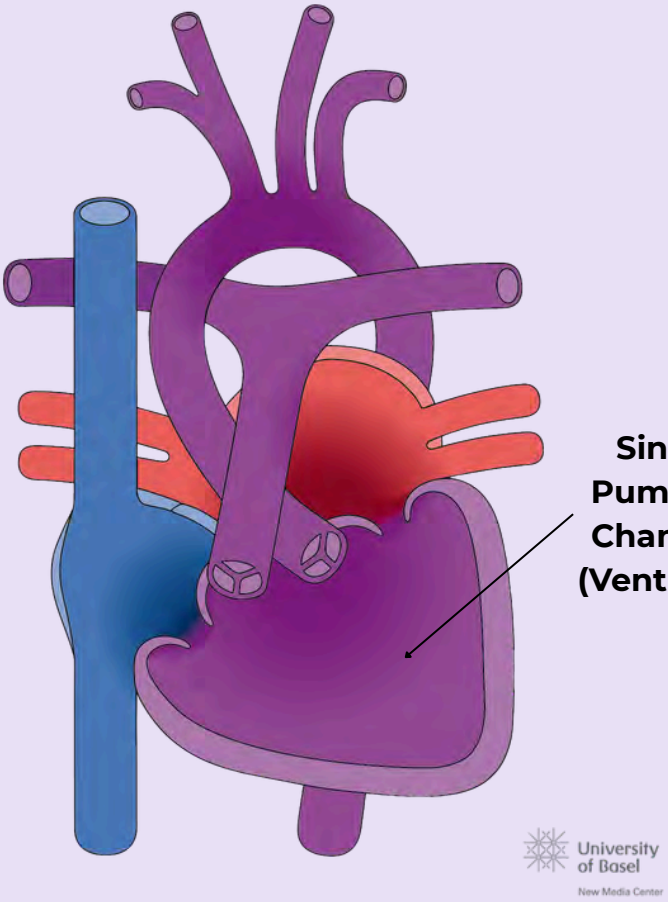
If you have Shone's complex with obstruction to left-sided heart structures and are planning a pregnancy, you may require a procedure to repair it prior to pregnancy. Talk to your ACHD doctor early in your planning process.

WHAT IS A SINGLE VENTRICLE?
(UNIVENTRICLE / FONTAN)

Structural Issues

A single ventricle refers to a group of conditions where one of the pumping chambers of your heart (either the right ventricle or the left ventricle) is not well developed (small in size) at birth.

There is only **one effective pumping chamber** (ventricle).



EXAMPLE: DOUBLE INLET LEFT VENTRICLE

SINGLE VENTRICLE: WHAT TO KNOW

What happens with single ventricle?

There is **complete mixing** of less oxygenated blood with oxygenated blood. The **single ventricle** has to now **pump** this mixed blood to the **lungs and the body**.

The body is unable to receive the normal amount of oxygen that it needs. To separate the less oxygenated and oxygenated blood, you **need a series of surgeries**.

Statistics

Single ventricle defects occur in about **1 in 15,000** live births, making up **less than 1%** of CHD cases.

It is estimated that ~70, 000 patients are living with a Fontan surgery worldwide; these numbers may double in the next 20 years.

Common Types of Single Ventricle Defects

Tricuspid Atresia: The tricuspid valve is very small or absent, which leads to an underdeveloped right ventricle and pulmonary artery.

Hypoplastic Left Heart Syndrome (HLHS): The mitral or aortic valve is very small or absent, which leads to an underdeveloped left ventricle and aorta.

Double Inlet Left Ventricle (DILV): Both upper chambers (atria) connect directly to a single left ventricle, while the right ventricle is very small or absent.

Unbalanced Atrioventricular Septal Defect (AVSD): The mitral and tricuspid valves do not form normally (valve may be narrow). This leads to reduced blood flow from the upper chambers (atria) to the lower chambers (ventricles), making one side of the heart much smaller than the other ("Unbalanced" AVSD). Balanced AVSD is where both ventricles are similar in size.

Common Types of Single Ventricle Defects

Pulmonary Atresia with Intact Ventricular Septum (PA/IVS): The pulmonary valve may be very small or absent, leading to an underdeveloped right ventricle and a narrowed or abnormal tricuspid valve.

Heterotaxy Syndrome: The heart and other organs are arranged differently in the body; one of the ventricles is often underdeveloped.

Not all CHD conditions that are associated with single ventricle are listed here (i.e., DORV, transposition)

SINGLE VENTRICLE: REPAIR STAGE 1

Treatment often involves a **series of surgeries** performed in **stages**, ultimately leading to a **Fontan procedure**. These are done to separate the less oxygenated from the more oxygenated blood and to decrease the heart's workload.

Most people with a single ventricle will have several procedures, but it is dependent on the individual.

Procedures may be performed in newborns to stabilize blood circulation to the lungs and body. The specific procedure is determined by the body's most urgent circulatory need (e.g., too much or too little blood flow to the lungs).

Catheter Procedures

A stent is placed in the patent ductus arteriosus to allow for blood to flow to the lungs or aorta, depending on which ventricle is smaller.

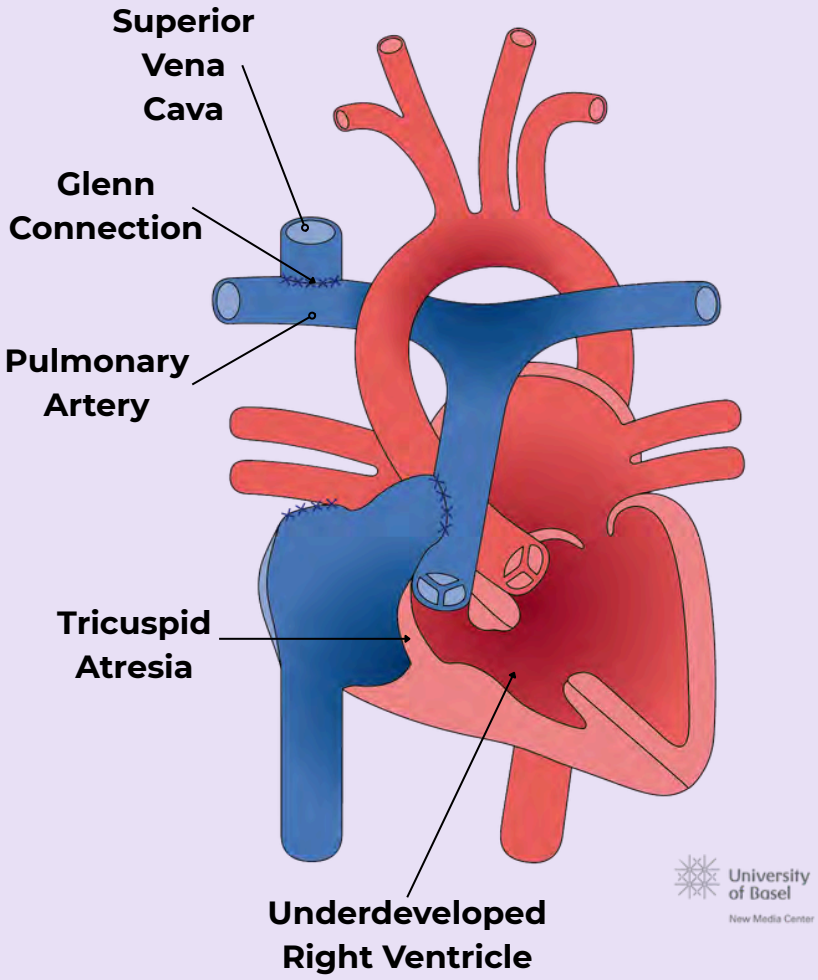
Surgical Procedures

Shunts (BTT shunt, Sano shunt, central shunt) may be used to increase blood flow to the lungs.
Norwood or DKS procedure may be done to enlarge the aorta.
Pulmonary artery banding may be done to limit too much blood flow to the lungs.

SINGLE VENTRICLE:
REPAIR STAGE 2

Surgical Procedure

Glenn procedure: Typically performed around 6 months after birth to direct less oxygenated blood from the upper body (from the superior vena cava) directly to the pulmonary artery (to the lungs), instead of going through the heart chambers (atria and ventricles). After this procedure, the less oxygenated blood from the upper body no longer mixes with the oxygenated blood in the heart.

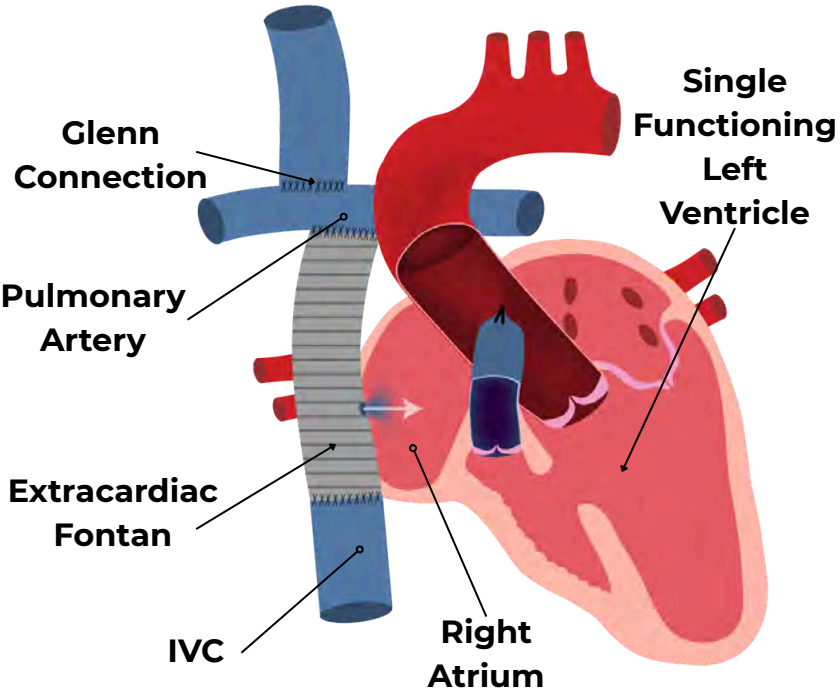


SINGLE VENTRICLE: REPAIR STAGE 3

Surgical Procedures

Fontan Procedure: Typically performed around 2-4 years old to direct less oxygenated blood from the lower body (from the inferior vena cava (IVC)) directly to the pulmonary artery. This reduces blood mixing, and oxygen levels in the body typically return to near-normal levels. Types of Fontan procedures include:

- The **IVC to pulmonary artery** connection could be completed using either the right atrium (lateral tunnel) or a tube outside the heart (extracardiac Fontan).



- **"Classic" type** (atrio/pulmonary Fontan) is an older, less commonly used technique in which blood is directed from the right atrium (carrying blood from the upper and lower body) directly to the pulmonary artery.

SINGLE VENTRICLE: LONG-TERM

All patients with a single ventricle, whether or not you underwent a Fontan procedure, need lifelong monitoring by an ACHD specialist to look out for:

***Electrical Issues*:** Abnormal heart rhythms, such as a **fast rhythm** (atrial flutter or ventricular tachycardia) or a **slow rhythm** (sinus node dysfunction), may develop due to surgical scars or stretching of the heart chambers over time. Treatment may include medications to control the rhythm or a pacemaker.

***Pumping Issues*:** Over time, the single ventricle may enlarge or weaken, making it harder for the heart to pump blood effectively. This can lead to fluid buildup in the lungs, abdomen, or legs. Medications may be needed to support heart function and reduce excess fluid buildup.

Not everyone will experience all long-term complications or symptoms.

SINGLE VENTRICLE: LONG-TERM

All patients with a single ventricle, whether or not you underwent a Fontan procedure, need lifelong monitoring by an ACHD specialist to look out for:

***Valve Issues*:** Heart valves may become leaky (regurgitation), which is more common, or narrow (stenosis), adding extra strain on the heart over time. Medications or surgery may be needed.

***Structural Issues*:** Narrowing can develop at various sites in the Fontan circulation. Pressure can also build up in the lungs or the Fontan pathway. Medications or surgery may be needed to lower pressures and improve blood flow.

Low Oxygen Levels: Some patients may develop bluish coloring of the lips or skin, called cyanosis. This can happen due to a small opening in the Fontan pathway (Fontan fenestration) or from extra blood vessels (collaterals) that can form in the lungs.

Not everyone will experience all long-term complications or symptoms.

SINGLE VENTRICLE: LONG-TERM

Blood Clots: Blood flows slower through the Fontan pathway, increasing the risk of blood clots. Clots can sometimes travel to the lungs (pulmonary embolism), brain (stroke), or other organs.

Medication Management

Over-the-counter medications such as aspirin, or prescribed blood thinners (warfarin, apixaban) may be used to reduce clotting risk or to treat blood clots.

Liver Problems: Because blood flow to the lungs in a Fontan circulation is passive (not pumped by the heart), pressure can build up, placing long-term stress on the liver.

- Over time, this may cause **scarring** (cirrhosis), which can lead to **complications** such as portal hypertension (high pressure in blood vessels of the abdomen), enlarged spleen, enlarged veins in the esophagus (varices), or bleeding from the gut.
- In rare cases, **advanced liver disease** can increase the risk of liver cancer, which is why regular monitoring is important.

Not everyone will experience all long-term complications or symptoms.

SINGLE VENTRICLE: LONG-TERM

Protein-Losing Enteropathy (PLE): Increased pressure in the Fontan circulation can affect the **lymphatic system** in the gut, causing **loss of proteins** (like albumin) into the stool. Low protein levels can lead to swelling in the legs, fluid around the lungs, or fluid in the belly.

Medication Management

Treatment may include medications such as steroids (budesonide) and blood thinners, along with a high-protein, low-fat diet.

Plastic Bronchitis: Abnormal lymphatic channels may leak protein-rich fluid into the airways. This can cause coughing and worsen oxygen levels. It may also cause sputum or mucus production.

Other Possible Issues:

- Varicose veins (prominent leg veins)
- Kidney problems
- Vitamin D deficiency
- Higher hemoglobin, especially if the oxygen in the blood is low for a long time
- Risk of infection involving the heart valves or Fontan pathway (endocarditis)

Not everyone will experience all long-term complications or symptoms.

SINGLE VENTRICLE: MANAGEMENT

Procedures to Restore Normal Heart Rhythm

- **Ablation:** A catheter procedure in which hot or cold energy is applied to restore the heart to a normal rhythm.
- **Cardioversion:** A brief electrical shock may be delivered to reset the heart back to normal rhythm. May be used if there is atrial flutter, atrial fibrillation, or ventricular tachycardia.
- **Pacemaker/defibrillator device** may be placed to address a slow or fast rhythm. The device is typically placed in your abdomen via a surgical procedure, and the wires are connected to the outside of the heart (also known as an **epicardial pacemaker**).

SINGLE VENTRICLE: MANAGEMENT

Interventional cath: If narrowing develops in the Fontan tube (conduit), a balloon or stent (metal mesh scaffold) can be used to keep it open.

Surgical repair: Surgery might be needed to treat any significant valve problem or, rarely, to change the Fontan type (e.g., from atrio/pulmonary to extracardiac type)

Transplant: If there is concern that the Fontan pathway is not working well or your heart/liver aren't functioning well, your doctor will assess for the potential need for a heart or combined heart and liver transplant.

Diagnostic Procedures

Liver Biopsy: This may be needed if there is concern for liver cirrhosis (scar tissue in the liver).

Diagnostic Cath: Depending on your symptoms and signs, a catheter procedure may be performed to assess the heart anatomy and pressures.

SINGLE VENTRICLE: COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid buildup in the lungs, legs, or belly.

Irregular heartbeats: Feeling your heart racing or skipping heartbeats (palpitations).

Abdominal symptoms: Bloating, pain, diarrhea, or poor appetite.

Infection: If you have persistent fever, flu-like symptoms, and/or night sweats, you should talk to your doctor to see if you have an infection that could be endocarditis.

Why do these things happen?

Tiredness: Over time, the single ventricle function might decline or the pressure in the blood vessels in your lungs might go up. This makes it harder for your blood to pick up oxygen or deliver it to the body, leaving you feeling exhausted even after mild activity.

Swelling: Can occur due to low protein levels, weak heart function, leaky valves, or high lung pressure.

Abdominal symtoms: More common when liver starts to be affected and become normal

Not everyone will experience all long-term complications or symptoms.

TIPS TO OPTIMIZE YOUR HEALTH

Stay well hydrated: Good hydration helps maintain blood flow through the Fontan pathway and reduces the risk of thickened blood or clots.

Do regular strength training: It is important to keep lower extremity muscles strong. This helps to pump the blood forward and improve circulation through the Fontan.

Focus on lung health: Effective blood flow through the Fontan depends on healthy lungs. Avoid inhalational agents (smoking, pollution) and do regular breathing practices (diaphragmatic/belly breathing).

FONTAN & PREGNANCY

Since Fontan circulation is unique, **pregnancy requires careful planning** and specialized counseling with an Adult Congenital Heart Disease (ACHD) team.

Why does pregnancy need to be carefully planned?

Pregnancy causes major changes in the body, including increased blood volume and increased workload on the heart. In Fontan circulation, the heart and blood flow may not tolerate these changes well.

There is currently limited long-term data. Despite uncertainties, more women with Fontan physiology are experiencing successful pregnancies, but they require careful planning and individualized considerations.

Risks to Mother

During pregnancy, some may develop irregular heart rhythms, worsening heart function, blood clots, or miscarriage.

Risks to Baby

Premature delivery and/or low birth weight are common.

SINGLE VENTRICLE:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits, ECG/EKG, Echocardiogram:
every year

Holter Monitor, Exercise Test:
every 1 - 2 years

Cardiac CT/MRI:
every 2 - 3 years

Heart Cath:
As indicated or once every 10 years

Labs:
every 1 - 2 years
(to assess blood cell count, hemoglobin, electrolyte levels, liver function, kidney function, cardiac markers, & stool studies to assess for protein leak)

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

WHAT IS TETRALOGY OF FALLOT?
(PRONOUNCED FAH-LOH)

Structural Issues

Tetralogy of Fallot is a combination of 4 ("tetra") CHD defects

1. **Pulmonary Stenosis or Atresia:** A narrowing (stenosis) or complete blockage (atresia) in the pathway from the right ventricle through the pulmonary artery to the lungs. This area is called the right ventricular outflow tract (RVOT).

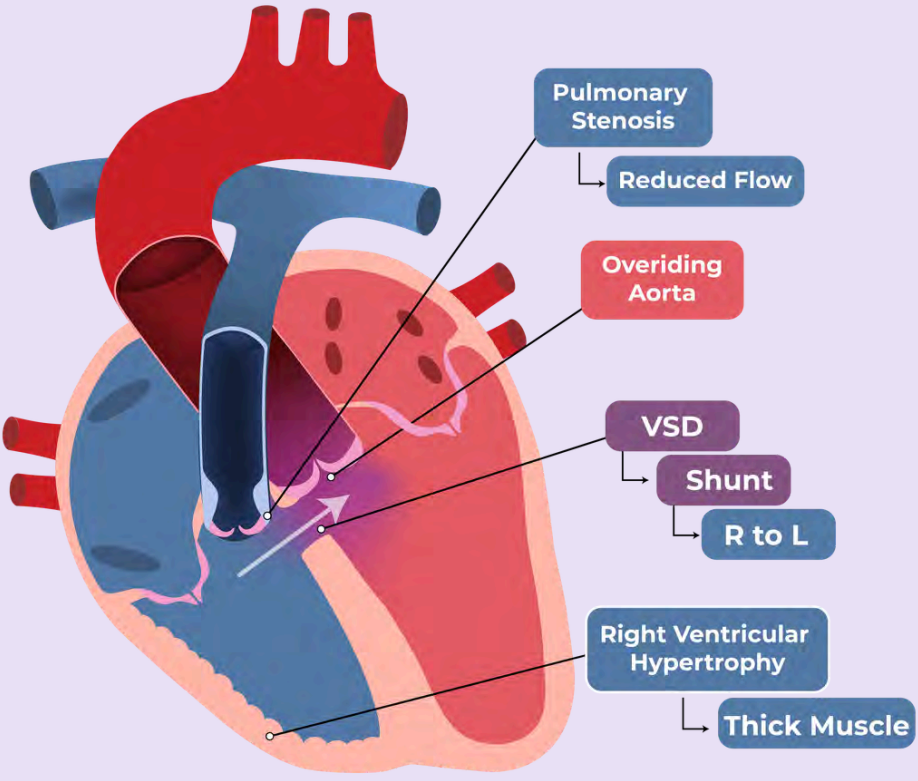
2. **Ventricular Septal Defect (VSD):** A hole in the wall (septum) between the two lower chambers (left and right ventricles) of the heart.

3. **Right Ventricular Hypertrophy (RVH):** The muscle of the right lower chamber (right ventricle) is thickened from working hard to pump blood through the narrowed or blocked pathway.

4. **Overriding Aorta:** The aorta is shifted and positioned over the VSD instead of arising completely from the left ventricle.

WHAT IS TETRALOGY OF FALLOT?

1. **Pulmonary Stenosis or Atresia**
2. **Ventricular Septal Defect (VSD)**
3. **Right Ventricular Hypertrophy (RVH)**
4. **Overriding Aorta**



TETRALOGY OF FALLOT

What Happens?

1. The narrowing caused by pulmonary stenosis **limits blood flow** to the **lungs**, so less blood can pick up oxygen.
2. Pulmonary stenosis also **raises the pressure** in the **right ventricle.** This high pressure pushes blood across the VSD from the right (R) to the left (L) ventricle, allowing less oxygenated **blood to mix** with more oxygenated blood
3. This mixing of blood **lowers oxygen levels**, which can cause cyanosis—a bluish color of the skin, lips, or nails.
4. Because the **right side of the heart** is **working harder** to push blood against the narrow pulmonary artery, its muscle becomes thicker over time (right ventricular hypertrophy).

TETRALOGY OF FALLOT

Statistics

Tetralogy of Fallot is a common cyanotic (blue) congenital heart disease, accounting for **5 - 7%** of CHDs.

It can be associated with genetic conditions such as *22q11.2* deletion (*~15%*).

If you have Tetralogy of Fallot, there is a slightly higher chance that congenital heart disease may occur in your close family members.

- Siblings: 2 – 3% chance of being born with CHD
- Children: 2 - 5% chance of being born with CHD

These risks may be higher if you also have an underlying genetic condition (such as *22q11.2* deletion syndrome or another chromosomal abnormality)

TETRALOGY OF FALLOT: EARLY PROCEDURES TO IMPROVE BLOOD FLOW

The approach to repairing Tetralogy of Fallot has changed over time and **varies depending on when and where** the surgery was done.

Some patients first had a temporary (palliative) procedure to improve blood flow to the lungs before complete repair, while others had full repair early in life.

Surgical Procedure

Blalock–Taussig–Thomas (BTT) Shunt: To increase blood flow to the lungs, a connection is made between the subclavian artery (a branch of the aorta that supplies blood to the arm) and the pulmonary artery. This connection is created by using the subclavian artery itself (classic BTT shunt) or, more commonly, using a synthetic tube (modified BTT shunt).

Catheter Procedures

In some cases, a **balloon** or **stent** is placed across the pulmonary stenosis or the patent ductus arteriosus (PDA), respectively, to help improve blood flow to the lungs **until full surgical repair** can be done.

TETRALOGY OF FALLOT: MORE COMPLETE REPAIR PROCEDURES

Surgical Procedures

Transannular Patch: A patch is placed across the pulmonary valve to enlarge the narrowed pulmonary valve pathway, allowing more blood to flow to the lungs.

VSD Closure: A patch is used to close the hole (VSD) between the lower chambers of the heart (ventricles). This prevents oxygen-rich and oxygen-poor blood fom mixing

Pulmonary Artery Plasty (PA plasty): If the right or left pulmonary arteries (PAs) are narrowed, a patch (PA plasty) may be placed during surgical repair to widen the PAs.

Conduit Placement: In some cases (e.g., if pulmonary atresia), a tube containing a valve (called a conduit) is surgically placed to connect the right ventricle to the pulmonary artery. This creates a new pathway for blood to flow to the lungs.

In some cases, pulmonary artery plasty may be completed via a catheter procedure.

TETRALOGY OF FALLOT: LONG-TERM

Not everyone will experience these long-term complications, but it is important to be aware of potential issues that may develop over time.

Valve Issues: One of the main long-term concerns after a complete initial repair is abnormal pulmonary valve function.

Valve Regurgitation: The transannular patch placed to relieve the pulmonary stenosis often prevents the valve from closing properly, allowing blood to leak backward into the right ventricle — also known as pulmonary valve regurgitation.

Residual or New Pulmonary Stenosis (Narrowing): Sometimes the narrowing may remain after surgery or a new narrowing may develop over time, particularly within the conduit placed in the right ventricular outflow tract (RVOT).

***Pump Issues*:** The valve problems can strain the right ventricle, causing it to become thickened (from stenosis) or enlarged (from regurgitation). Over time, this strain can weaken the right ventricle's pumping ability, and eventually it may also affect the left ventricle's pumping function.

TETRALOGY OF FALLOT: LONG-TERM

Not everyone will experience these long-term complications, but it is important to be aware of potential issues that may develop over time.

Electrical Issues: Scar tissue from surgery can disrupt the heart's electrical system, leading to **abnormal fast heart rhythms (arrhythmias)**. Examples include atrial flutter, atrial fibrillation, and ventricular tachycardia.

Structural Issues: The **aorta** may enlarge over time and require close monitoring. The **right or left pulmonary arteries** may become narrow over time (called as branch pulmonary artery stenosis).

***Infection Risk*:** If your pulmonary valve has been replaced, there is a risk of infection in the valve or inner lining of the heart (endocarditis).

TETRALOGY OF FALLOT: COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Chest pain, shortness of breath: Especially with exercise.

Why do these things happen?

If your **pulmonary valve isn't working** well because of narrowing or leakage, you may have no symptoms, or you may feel tired, short of breath, or have chest pain, especially during exercise.

If the right side of your heart has **decreased pumping function,** you may experience **swelling** in your legs or abdomen.

You may feel **palpitations**, such as your heart racing or skipping beats, due to an abnormal fast heart rhythm. Sometimes this can cause **dizziness** or **fainting**.

Not everyone will experience all long-term complications or symptoms.

TETRALOGY OF FALLOT: LONG-TERM MANAGEMENT

Pulmonary Valve Replacement: Depending on the severity of pulmonary valve narrowing or leakage, the size of your right ventricle, and symptoms, your doctor may recommend placing a new pulmonary valve. This can often be done with a catheter procedure, but sometimes requires surgery depending on your anatomy.

Pulmonary Artery Stenting: If narrowing develops in the right or left pulmonary arteries over time, a metal mesh tube (stent) can be placed via a catheter to open up the narrowed area and improve blood flow to the lungs.

Arrhythmia Procedures:

Pacemaker/ Defibrillator (ICD): A device may be placed under the skin to protect against serious, life-threatening arrhythmias by monitoring your heart rhythm and delivering treatment when needed.

Ablation: A catheter procedure to eliminate the source of abnormal rhythms and restore normal heart rhythm.

Cardioversion: A brief electrical shock to reset the heart rhythm if you develop atrial flutter, atrial fibrillation, or ventricular tachycardia.

TETRALOGY OF FALLOT: LONG-TERM MEDICAL MANAGEMENT

Medication Management

Depending on your specific condition and symptoms, you may need one or more of the following medications:

Heart Rhythm Medications: Medications to control fast or irregular heart rhythms (arrhythmias) and help maintain a normal heart rate.

Blood Thinners (Anticoagulation): You may need to take blood thinners (such as warfarin, apixaban, or other newer anticoagulants) if you have a history of irregular heart rhythms such as atrial fibrillation. This helps lower your risk of stroke and blood clots.

Medications to Improve Heart Pumping Function: If your heart's pumping function has weakened, you may need medications that help your heart pump more effectively and reduce strain on the heart. These may include ACE inhibitors, beta-blockers, or diuretics (water pills).

TETRALOGY OF FALLOT:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every 1 - 2 years

Exercise Test:
every 2 - 5 years

Heart Monitor:
as needed

Cardiac CT/MRI:
every 2 - 3 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

Transposition of the Great Arteries (TGA)

Transposition of the Great Arteries is a condition where both the **great arteries** are **switched**.

There are **2 types** of TGA:

- **D-TGA:** Dextro-transposition of the great arteries
- **CC-TGA:** Congenitally-corrected transposition of the great arteries **also known as **L-TGA** (Levo-transposition of the great arteries) **or ventricular inversion**"*

We will first go over **D-TGA** and then **CC-TGA.**

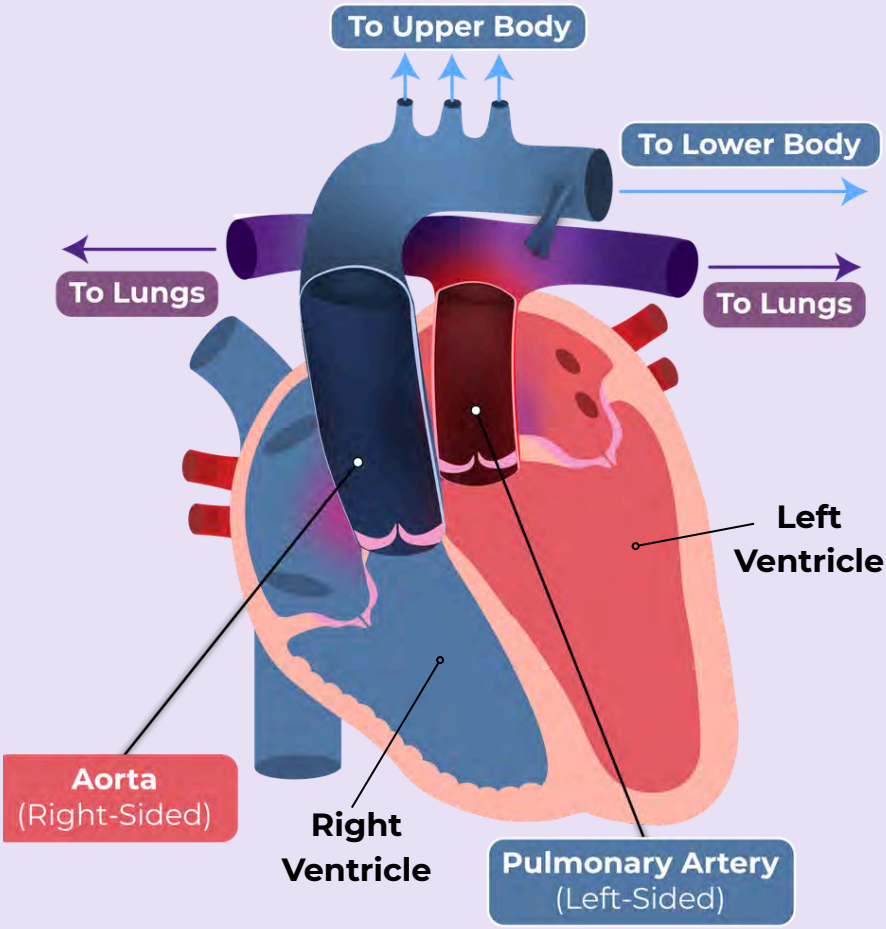
If you would like to skip directly to CC-TGA, click through to **page 171.**

WHAT IS A D-TGA?

Structural

Dextro-transposition of the great arteries (D-TGA) is a condition in which the two main arteries leaving the heart are **switched from their normal positions**

- Normally, the pulmonary artery connects to the right ventricle and carries oxygen-poor blood to the lungs. The aorta connects to the left ventricle and carries oxygen-rich blood to the body.
- In D-TGA, the pulmonary artery is abnormally connected to the left ventricle and the aorta to the right ventricle.



WHAT IS D-TGA?

What Happens?

- **Less-oxygenated blood** returning from the body enters the right ventricle, which then pumps it through the aorta back out to the body, without ever going to the lungs to pick up oxygen.

- At the same time, **oxygenated blood** from the lungs goes to the left ventricle, which pumps it through the pulmonary artery back to the lungs, where it picks up more oxygen that it doesn't need.

- This creates **two separate,** or **“parallel,” circulation loops** instead of a single connected system. This means the body keeps receiving less oxygenated blood and never gets the oxygen it needs. The lungs keep receiving oxygenated blood that should be going to the body

- Because the body receives very little oxygen, babies with D-TGA develop a **bluish discoloration** (cyanosis) of the skin and lips shortly after birth.

D-TGA:
WHAT TO KNOW

Statistics

D-TGA makes up about **3%** of CHDs

Associated Conditions:

Structural Issues

Ventricular Septal Defect (VSD): A hole between the lower chambers of the heart.

Pulmonary Valve Stenosis: The pulmonary valve may be narrowed.

Coarctation of Aorta or Interrupted Aortic Arch: The aorta may not form normally. Coarctation of the aorta is a narrowing of the artery. Interruption of the aortic arch results in severe narrowing of the artery.

Pulmonary Artery Stenosis: The arteries to the lungs may be narrowed.

Coronary Artery Anomalies: The arteries (coronaries) that supply blood to the heart muscle may arise from a different location in the aorta.

D-TGA:
PRIMARY REPAIRS

Surgery is needed for repair and is typically performed in the first weeks of life. Your long-term health and follow-up care depend on which type of repair you had.

Depending on when you were born and the specifics of your anatomy, you may have undergone one of the following procedures:

Atrial Switch Procedure (Mustard or Senning operation) - commonly performed before the 1990s
Arterial Switch Operation- the preferred procedure since the 1990s and most common today
Rastelli Procedure - used for D-TGA with additional defects like VSD and pulmonary stenosis

Catheter Procedure

Balloon Atrial Septostomy: Creates or enlarges a hole between the upper chambers of the heart (atria), allowing oxygenated and less oxygenated blood to mix. This temporarily improves oxygen levels in babies before definitive surgery can be performed.

D-TGA:
SURGICAL REPAIRS

Other Surgical Procedures

Réparation à l'Etage Ventriculaire (REV) Procedure: A complex repair that creates a tunnel inside the heart to redirect blood flow from the left ventricle through the VSD to the aorta, while also connecting the pulmonary artery to the right ventricle with a conduit. This is used for D-TGA with VSD and pulmonary stenosis.

Nikaidoh Procedure: A specialized type of arterial switch operation used when there is also narrowing below the aortic valve. It involves moving the aorta to its correct position over the left ventricle, relocating the coronary arteries, and reconstructing the right ventricular outflow tract (RVOT) using the patient's own tissue.

Now, we will review each of the primary repair procedures in detail in the following order:

1. **Atrial Switch Procedure**
2. **Arterial Switch Operation**
3. **Rastelli Procedure**

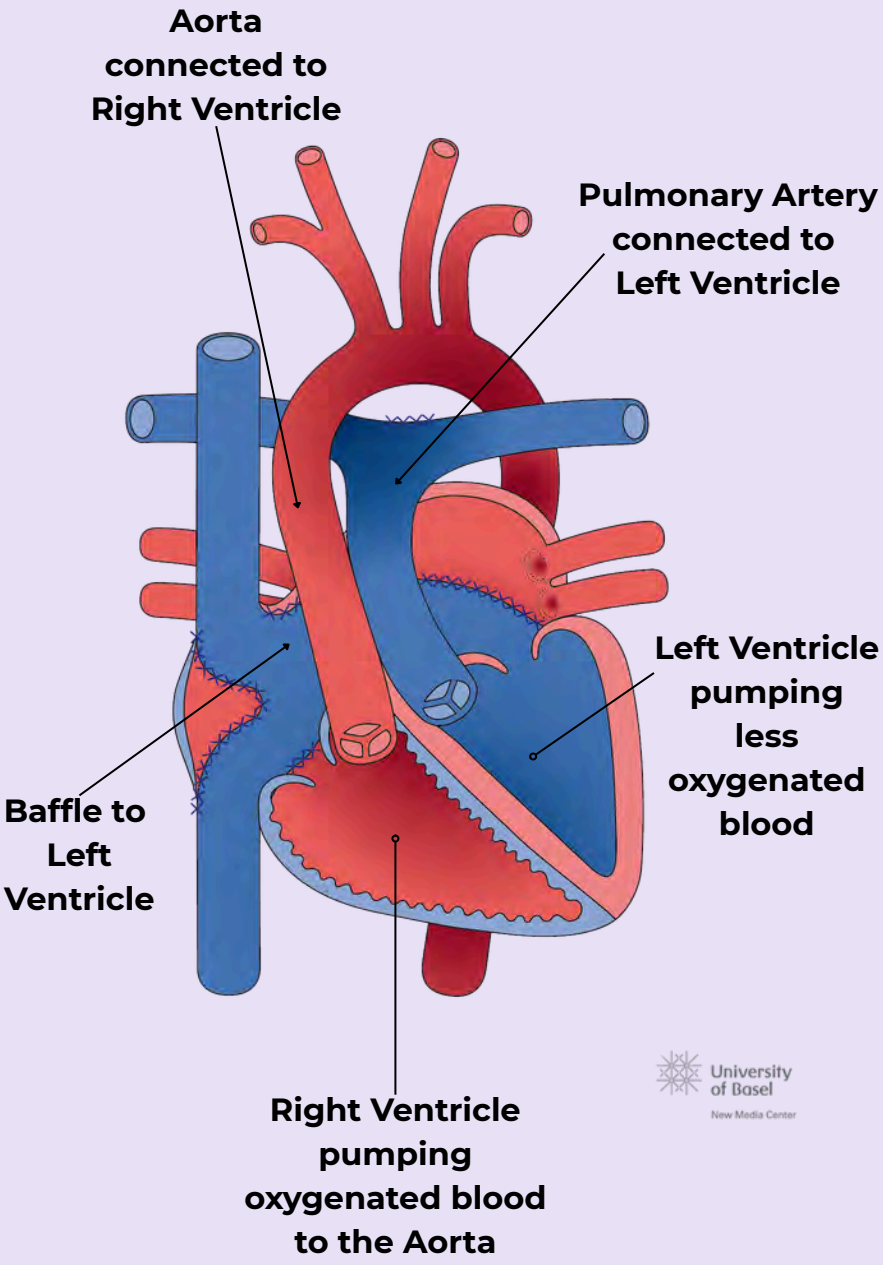
1 ATRIAL SWITCH PROCEDURE

MUSTARD / SENNING

- Common **before the 1990s**.
- Performed using a **Mustard** or **Senning** procedure, which both use a similar approach to atrial switch.
- The procedure redirects blood flow at the atrial (upper chamber) level using **baffles** (internal tunnels):
  - Less oxygenated blood returning from the body is redirected to the **left ventricle, which pumps it to the lungs** through the pulmonary artery.
  - Oxygenated blood from the lungs is redirected to the **right ventricle, which pumps it to the body** through the aorta.
- This procedure reroutes the blood but does not switch the great arteries back to their normal positions. The right ventricle remains connected to the aorta, meaning it must pump blood to the entire body for life—a job it was not designed to do.

1 ATRIAL SWITCH PROCEDURE

MUSTARD / SENNING

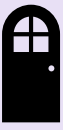


1 D- TGA *AFTER* ***ATRIAL SWITCH*****:** LONG-TERM

Structural Issues: Narrowing (baffle stenosis) or leakiness (regurgitation) from the patch/pathway created in the atria. This can increase the risk of blood clot formation and stroke.

Pump Issues: The right ventricle may become enlarged (dilated) and have a decreased ability to pump blood effectively to the aorta. This may lead to increased fluid in the lungs or body (heart failure) or high pressure in the lungs (pulmonary hypertension).

Electrical Issues: Abnormal heart rhythms due to scarring in the atria. This could be a fast rhythm (atrial flutter, atrial fibrillation) or a slow rhythm (sinus node dysfunction). A fast heart rhythm originating in the lower chambers (ventricular tachycardia) can also occur, particularly when the right ventricle's pumping function is decreased.



Valve Issues: Heart valves (commonly the tricuspid valve) may become leaky (regurgitant) as the right ventricle dilates and stretches the tricuspid valve.

Not everyone will experience all long-term complications or symptoms.

1 D- TGA *AFTER* ***ATRIAL SWITCH*****:** COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Shortness of breath, chest pain: especially with activity.

Why do these things happen?

Tiredness: If blood pressure in your lungs becomes too high, it may become hard for the blood to carry enough oxygen to the body. Your oxygen levels might decrease, and you might feel tired easily even with small activity.

Swelling: Can occur due to weak heart function, leaky valves, or high lung pressure.

Irregular heartbeats: **see "Electrical Issues"*

What are the potential risks?

Stroke: If a clot forms, it may travel to the brain and can cause a stroke.

Right Ventricle Failure: If the right ventricle becomes weak and symptoms persist despite medications, a heart transplant may be considered.

Not everyone will experience all long-term complications or symptoms.

1 D- TGA *AFTER* ***ATRIAL SWITCH*****:** MANAGEMENT

Surgical Procedure

Tricuspid Regurgitation: If the heart valve becomes leaky (regurgitation), a surgical procedure may be needed to repair or replace the valve.

Catheter Procedure

Arrhythmias: May need a procedure such as cardioversion or catheter ablation. In some cases, a defibrillator may be needed.

Slow Heart Rhythms: May require a pacemaker to help regulate the heartbeat.

Baffle narrowing (stenosis) or **leak** (regurgitation): If significant stenosis, a stent (mesh scaffold) may be needed. If significant leak, a small device can be used to close the leak.

Medication Management

Right Heart Failure: If the right ventricle has decreased pumping function, you may be placed on medications such as diuretics, ACE inhibitors, beta-blockers, or others.

2 ARTERIAL SWITCH OPERATION (ASO)

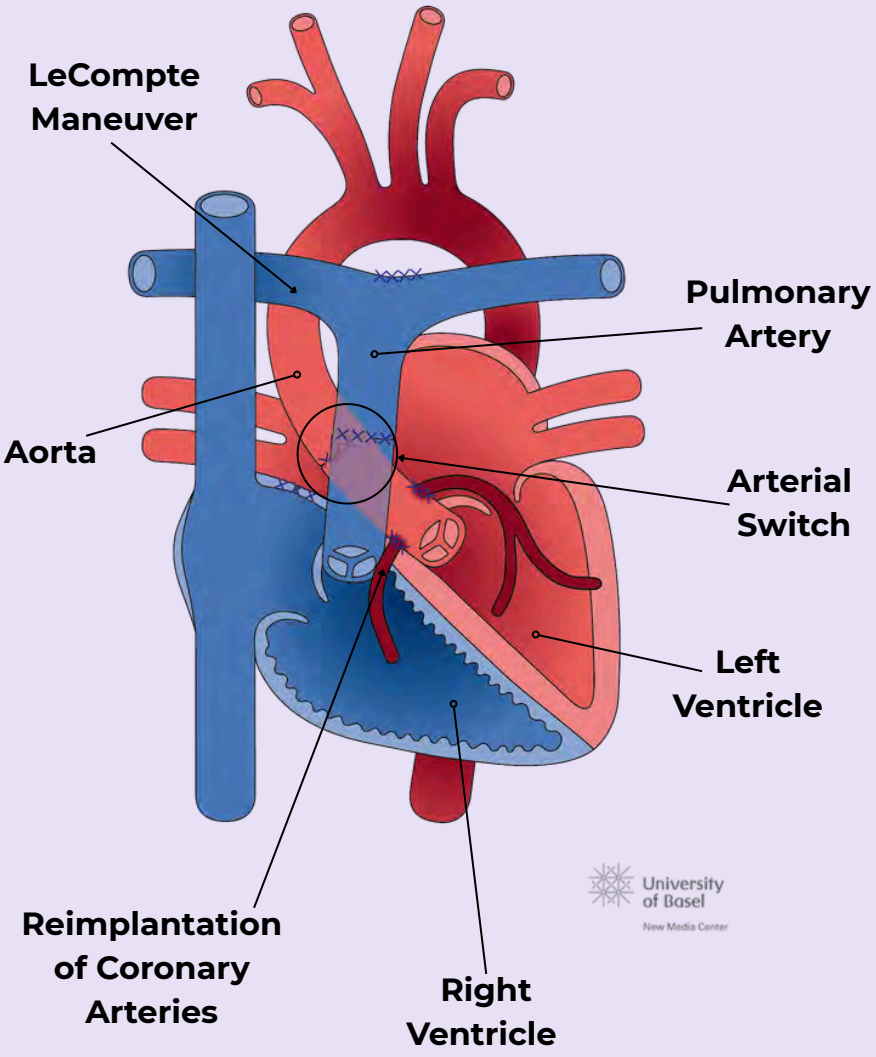
JATENE

- **More common** since the 1990s.
- Also known as the **Jatene** procedure.
- The surgery anatomically corrects the transposition by switching the great arteries to their normal positions: the aorta and pulmonary artery are disconnected near their origins; they are then reconnected to the correct pumping chambers (i.e., the aorta to the left ventricle and the pulmonary artery to the right ventricle).
- This restores normal circulation, with the left ventricle pumping to the body and the right ventricle pumping to the lungs
- **Key components of surgery include:**
  Lecompte Maneuver: During the arterial switch surgery, the pulmonary arteries are brought forward in front of the aorta so they can connect properly to the right ventricle.

  Coronary Re-Implantation: During this surgery, the coronary arteries, which supply blood to the heart muscle, are moved and reattached to the new aorta ("neo-aorta").

2 ARTERIAL SWITCH OPERATION (ASO)

JATENE



2 D- TGA *AFTER* ***ARTERIAL SWITCH***: LONG-TERM

Structural Issues: Narrowing (stenosis) can develop at the surgical connection sites of the aorta or pulmonary artery, which may affect blood flow.

- **Pulmonary artery stenosis:** Narrowing of the arteries that take blood to the lungs. This happens as the arteries are pulled forward during surgical repair to connect to the right ventricle.

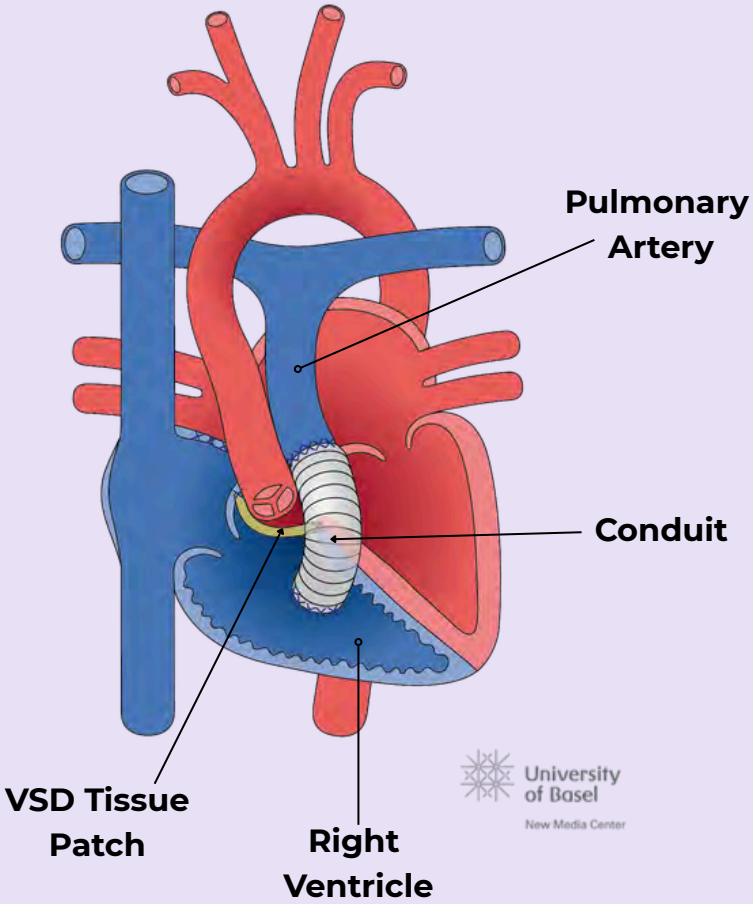
- **Coronary artery stenosis**: As the coronary arteries are relocated to the new aorta (neo-aorta), scarring at the site of connection may narrow the vessels.

Valve Issues: The heart valves (the aortic and pulmonary valves) may develop leakiness (regurgitation) over time.

Not everyone will experience all long-term complications or symptoms.

3 RASTELLI PROCEDURE

- The Rastelli procedure is **used when there is a hole** between the lower pumping chambers (VSD) and narrowing of the pulmonary valve (stenosis).
- Involves **placing a tube with a valve (conduit)** to direct blood from the right ventricle to the lungs.
- The hole (VSD) is closed with a **tissue patch,** directing the blood from the left ventricle to the aorta.



3 D- TGA *AFTER* ***RASTELLI*****:** LONG-TERM

Structural Issues: The RV-PA conduit may become narrow (stenosis) or leak (regurgitation), requiring surgical replacement of the tube (conduit). A prosthetic pulmonary valve (Melody valve or Harmony valve) may be placed via a catheter procedure. There may also be narrowing of the pathway from the left ventricle to the aorta (subaortic stenosis) created by closing the VSD.

Valve Issues: The aortic valve may develop narrowing (stenosis) or may leak (regurgitation) with time.

Electrical Issues: Heart rhythm problems can occur due to surgical scarring of heart tissue, including fast rhythms (atrial fibrillation, ventricular tachycardia) or slow rhythms (complete heart block).

Plumbing Issues**:** Pulmonary artery branches may develop narrowing (stenosis).

Infection Risk**:** Infection of the heart valves, the RV-PA conduit, or the heart lining (endocarditis).

Not everyone will experience all long-term complications or symptoms.

23 D- TGA *AFTER* ***ARTERIAL SWITCH / RASTELLI*****:** COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Shortness of breath, chest pain: especially with activity.

Why do these things happen?

Tiredness: If blood pressure in your lungs becomes too high, it may become hard for the blood to carry enough oxygen to the body. Your oxygen levels might decrease, and you might feel tired easily even with small activity.

Swelling: Can occur due to weak heart function, leaky valves, or high lung pressure.

Irregular heartbeats: **see "Electrical Issues"*

What are the potential risks?

Arterial switch: If the coronary arteries become narrow, not enough blood and oxygen can reach the heart. This can lead to a heart attack.

Rastelli: Risk of right ventricle failure when the RV-PA conduit develops severe narrowing or leaking. If symptoms persist, a heart transplant may be considered.

Not everyone will experience all long-term complications or symptoms.

23 D- TGA *AFTER* ***ARTERIAL SWITCH / RASTELLI*:** MANAGEMENT

Surgical Procedures

Valve Repair or Replacement: If the aortic or pulmonary valve becomes severely leaky, either repair or replacement may be necessary. If the neo-aorta is dilated (enlarged), it may need repair as well.

RV-PA Conduit Replacement: After a Rastelli procedure, you might need to repair or replace the conduit. **usually surgical, but can be done via catheter in some cases*

Coronary Artery Stenosis may require repair (can be done via catheter or surgery)

Catheter Procedures

Pulmonary Artery Stenosis: A balloon or stent (mesh scaffold) is placed to open up the artery.

Arrhythmias: May need a cardioversion or catheter ablation. In some cases, a defibrillator is needed.

Slow Heart Rhythms: May require a pacemaker to help regulate the heartbeat.

Medication Management

If the pumping function of the ventricles is decreased, medications may be needed.

1 2 3

D-TGA:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every year

Exercise Test:
every 3 - 5 years

Heart Monitor:
every 2 years (if you had an atrial switch)

Cardiac CT/MRI:
every 2 - 3 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS CC-TGA?

Before you get started, please note:

Congenitally-corrected transposition of the great arteries (CC-TGA), Levo-transposition of the great arteries (L-TGA), and ventricular inversion **are all different names for the same defect**. Here, we will use "CC-TGA".

If you have CC-TGA with a single ventricle, the separate single ventricle section will be more applicable to you.

If you have CC-TGA but not single ventricle, proceed with this section.

You will be able to receive a PDF of all the conditions at the end.

Structural Issues

CC-TGA is a condition where both the **heart's pumping chambers (ventricles) and the great arteries are switched**:

- The right ventricle is positioned on the left side and connects to the aorta (instead of the pulmonary artery)
- The left ventricle is positioned on the right side and connects to the pulmonary artery (instead of the aorta)

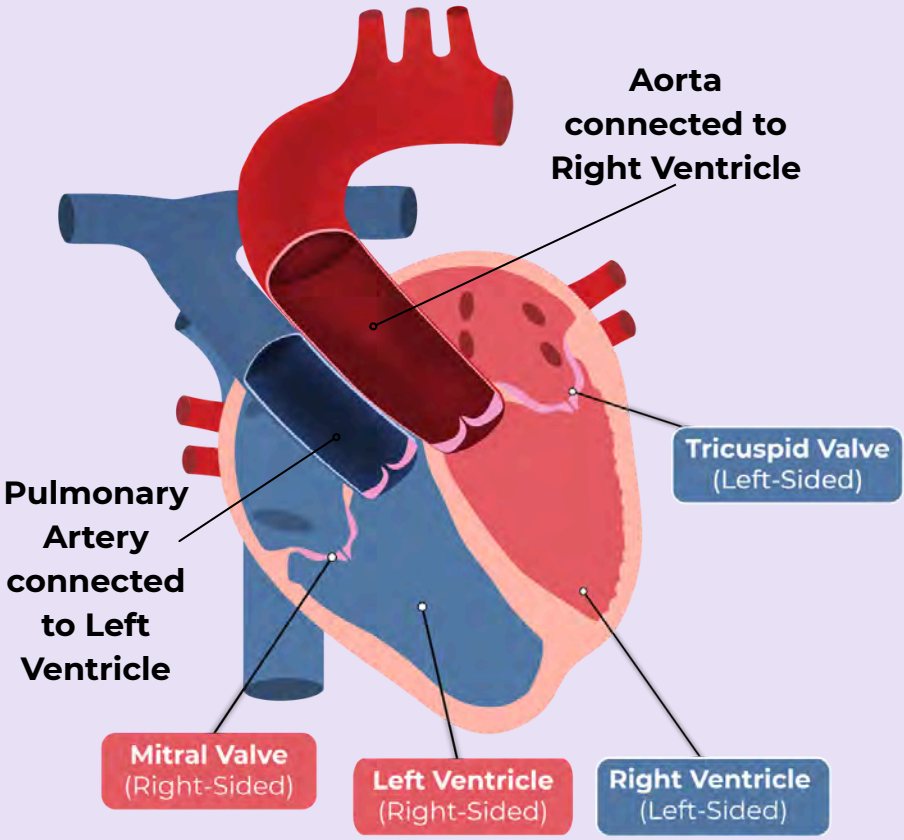
WHAT IS CC-TGA?

What Happens?

The **right ventricle receives oxygenated blood** from the lungs and pumps it to the body through the aorta, while the l**eft ventricle receives less oxygenated blood** from the body and pumps it to the lungs.

This **keeps oxygen levels normal**, which is why the condition is called congenitally-corrected transposition of the great arteries.

Since the right ventricle is not designed to pump against high pressure in the aorta, the right ventricle may become weaker with time.



CC-TGA:
WHAT TO KNOW

Statistics

CC-TGA is rare, occurring in **0.05%** of babies born with CHD.

Associated With:

Structural Issues

Ventricular Septal Defect (VSD): A hole between the two lower pumping chambers of the heart.

Pulmonary Stenosis: Narrowing of the pulmonary valve and/or pulmonary arteries, which limits blood flow to the lungs. Rarely, the pulmonary valve may be absent (pulmonary atresia)

Abnormal Tricuspid Valve: The tricuspid valve may be malformed (Ebstein anomaly) and can become leaky.

Slow Heart Rhythm: Problems with the heart's electrical system can cause a slow heartbeat, including complete heart block.

CC-TGA:
PHYSIOLOGIC REPAIR

There are **two main surgical approaches** for CC-TGA: “physiologic repair” and “anatomic repair”. Which repair is used depends on the individual’s heart anatomy and feasibility.

Surgical Procedure: Physiologic Repair

The **right ventricle remains the main pumping chamber** to the body. This improves symptoms and short-term blood flow. Surgery focuses on fixing associated heart problems, such as:

- Closing a ventricular septal defect (VSD) using a tissue patch.
- Widening the narrowed pulmonary valve area using a tissue patch and/or replacing it with an artificial valve.
- Repairing or replacing the tricuspid valve.

Risk: *In the long term, there is strain on the right ventricle. This may lead to heart failure.*

Some people may be diagnosed as adults, and some may not require surgery.

CC-TGA:
ANATOMIC REPAIR

There are **two main surgical approaches** for CC-TGA: “physiologic repair” and “anatomic repair”. Which repair is used depends on the individual’s heart anatomy and feasibility.

Surgical Procedure: Anatomic Repair

The goal is to make the **left ventricle the body's main pumping chamber** by connecting it to the aorta. The right ventricle is connected to the pulmonary artery. This is a “**double switch**” and reduces long-term strain on the right ventricle.

Risk: *A newer surgical strategy. It may not be suitable for all patients.*

CC-TGA:
LONG-TERM

Pump Issues: The right ventricle is not designed to pump blood against the high pressure in the aorta. This extra workload can cause the right ventricle to enlarge and/or weaken, leading to heart failure.

Valve Issues: Tricuspid valve insufficiency (leak) results in increased blood flow and strain on the right ventricle.

Electrical Issues:
Due to **abnormal electrical pathways**, there is a risk of a slow heart rate (example: complete heart block).

Due to **scar tissue or stretched heart chambers**, abnormal fast heart rhythms can occur, such as atrial fibrillation or ventricular tachycardia.

Some people may be first diagnosed as adults, and may not require surgery.

Not everyone will experience all long-term complications or symptoms.

CC-TGA:
COMMON SYMPTOMS

Possible Symptoms

Tiredness, Fatigue: Feeling more tired than usual.

Shortness of Breath, Chest Pain especially with activity.

Swelling: Extra fluid in your body.

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Slow Heart Rate: Lightheadedness, dizziness.

Why do these things happen?

Abnormal function of the right ventricle or tricuspid valve.

Abnormal electrical pathways, scar tissue, and stretched heart chambers.

Not everyone will experience all long-term complications or symptoms.

CC-TGA:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every year

Heart Monitor:
every 1 - 5 years

Exercise Test*, *Cardiac CT/MRI:
every 3 - 5 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendation may vary based on individual needs and provider preferences.*

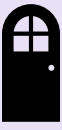
If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

TRICUSPID VALVE ANOMALIES

IS THIS SECTION FOR ME?

While this section focuses specifically on Ebstein Anomaly, it is designed for **anyone** diagnosed with a structural problem of the **tricuspid valve**.

Whether your tricuspid valve is "**displaced**" or "**shaped abnormally**", the result is often the same: *the valve leaks, and the right side of the heart works harder.*



Common Tricuspid Valve Issues

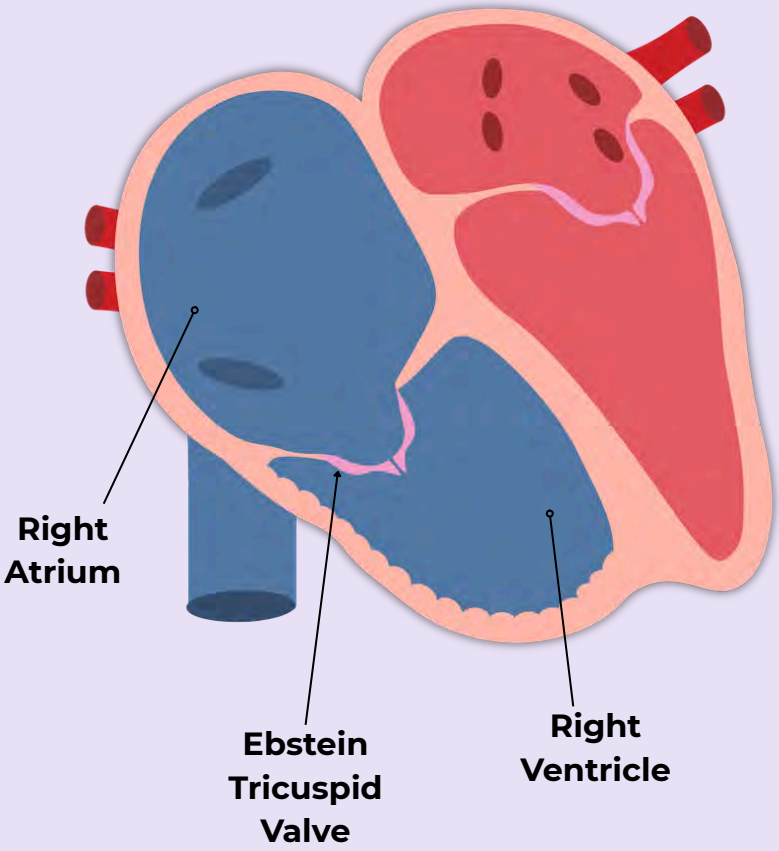
- Ebstein's Anomaly
- Tricuspid Valve Dysplasia
- Cleft Tricuspid Valve
- Severe Tricuspid Regurgitation

WHAT IS EBSTEIN ANOMALY?

Structural Issues

Displaced Valve: The tricuspid valve is abnormal in shape and sits lower in the heart than normal (displaced).

Chamber Size: This low position enlarges the top chamber (right atrium) and reduces the lower pumping chamber (right ventricle) to a smaller than usual size.



WHAT IS EBSTEIN ANOMALY?

Valve Issues

Leaky Valve: Due to its abnormal shape and position, the valve does not close properly. This allows blood to leak backward across the valve (regurgitation).

Pump Issues

Heart Strain: The severity of the leak varies, but it will lead to the enlargement of the right-sided heart chambers over time.

Diagnosis Timing: *Depending on the degree of leaking, you may be diagnosed early in life or later in adulthood.*

Statistics

Ebstein's Anomaly accounts for **less than 1%** of all types of congenital heart disease and is more common in twins.

EBSTEIN ANOMALY: WHAT TO KNOW

Associated With:

Structural Issues

Atrial Septal Defect (ASD): A hole between the upper chambers (atria).

Ventricular Septal Defect (VSD): A hole between the lower chambers (ventricles).

Pulmonary Stenosis: Narrowing of the pulmonary valve, which controls blood flow to the lungs.

Electrical Issues

Wolff-Parkinson-White (WPW) Syndrome: An extra electrical pathway that can cause very fast heartbeats.

EBSTEIN ANOMALY: COMMON PROCEDURES

If the tricuspid valve is mildly abnormal and there is only a mild valve leak (regurgitation), you might not need any procedure.

Surgical Procedures

Valve Repair: If there is severe valve leakiness or symptoms of heart failure, then surgical repair is needed. This may be a Cone procedure or replacement of the tricuspid valve.

Glenn Procedure: In some cases where the right ventricle's pumping function is significantly decreased, a Glenn procedure may be performed in combination with tricuspid valve repair. This procedure redirects blood flow from the upper body directly to the lungs, reducing the workload on the weakened right ventricle while still allowing it to pump to the body.

Catheter Procedures

An **ablation** may be performed to restore the heart to a normal rhythm if an arrhythmia is present.

Cardioversion may be used if there is atrial flutter, atrial fibrillation, or ventricular tachycardia.

EBSTEIN ANOMALY: LONG-TERM

Valve Issues: The tricuspid valve is misplaced and structurally abnormal, and it does not close well. If the valve does not close, blood can flow backwards (regurgitation), possibly causing enlargement of the right atrium. This increases the risk of abnormal heart rhythm.

***Pump Issues*:** When the tricuspid valve leaks, extra blood flows backward into the right ventricle, causing it to enlarge and weaken over time. The right ventricle muscle is not well developed. In combination with the valve issues, this can lead to decreased pumping function.

***Electrical Issues*:** Usually caused by scarring of heart chambers when they are enlarged or when there is extra electrical tissue in the heart. Abnormal heart rhythms may occur related to abnormal electrical pathways, and are seen in ~50% of people with Ebstein Anomaly.

Not everyone will experience all long-term complications or symptoms.

EBSTEIN ANOMALY: COMMON SYMPTOMS

Possible Symptoms

Tiredness, fatigue: Feeling more tired than usual.

Shortness of breath, chest pain especially with activity.

Swelling: Extra fluid in your body.

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations or arrhythmias).

Why do these things happen?

Reduced function of the right ventricle can cause heart failure symptoms such as shortness of breath and leg swelling.

What are the potential risks?

Stroke: If there is a hole in the wall of the heart, blood clots can pass through the hole and travel to the brain.

Not everyone will experience all long-term complications or symptoms.

EBSTEIN ANOMALY:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every 1 - 2 years

Heart Monitor:
as needed

Exercise Test, Cardiac CT/MRI:
every 2 - 3 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

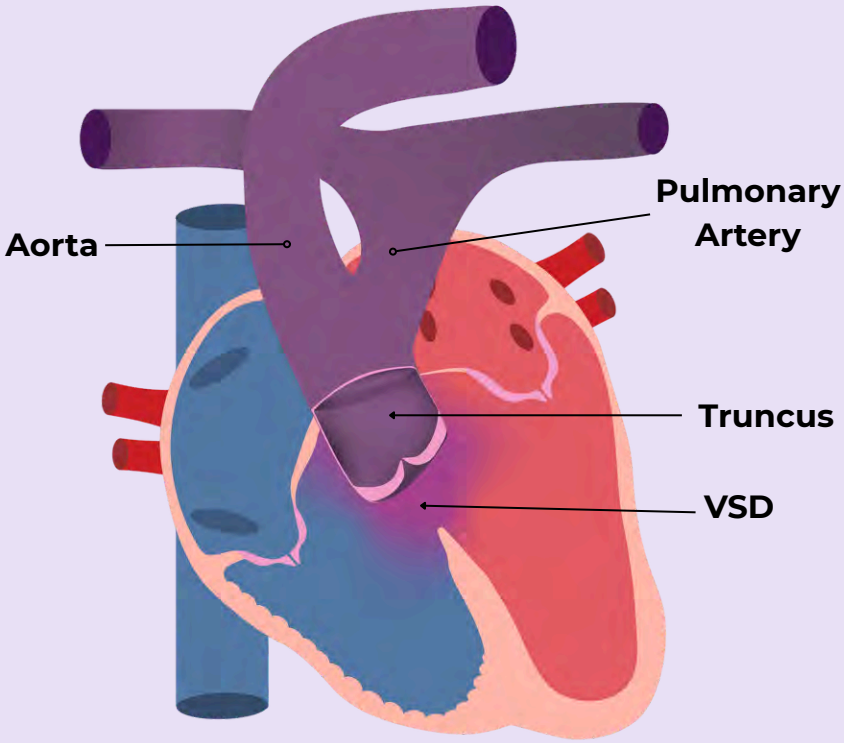
WHAT IS TRUNCUS ARTERIOSUS?

Structural Issues

Truncus arteriosus is a heart condition where there is a single large blood vessel coming out of the heart instead of having two blood vessels. This **single large blood vessel is** called the **“Truncus”**, which branches to the aorta and the pulmonary artery.

The truncus usually sits over a **hole** between the two lower chambers of the heart (called a **ventricular septal defect, or VSD**).

The truncus vessel has **one valve**, called the **truncal valve**. This valve may **leak** (regurgitation), be **narrow** (stenosis), or be a combination of both.



TRUNCUS ARTERIOSUS: WHAT TO KNOW

What Happens?

There is **mixing** of **less-oxygenated** and **oxygenated blood** in the **truncus** before it splits into the aorta and the pulmonary artery, carrying the mixed blood to both the lungs and the body.

Statistics

Truncus Arteriosus is a rare condition, making up l**ess than 1%** of all CHD.

Associated With:

Structural Issues

Interruption of the aortic arch (portion of the artery is missing) or **coarctation of the aorta** (narrowing of the artery).

It may also be associated with an **absent or narrow artery to the lung (pulmonary artery)** or **coronary artery anomalies.**

Genetics

22q11.2 deletion (DiGeorge syndrome)

TRUNCUS ARTERIOSUS: MANAGEMENT

Surgical repair is usually performed in the **first few weeks** of life to separate the circulation and establish normal blood flow to the lungs and body. This includes several steps:

Surgical Procedures

Blood flow to the lungs is separated from the blood flow to the body by **creating two different pathways**.

A tube (conduit) is placed in the heart **connecting the right ventricle to the pulmonary artery** (RV to PA conduit or homograft). This carries less oxygenated blood from the heart to the lungs.

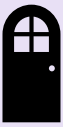
The hole (VSD) is closed with a **tissue patch**.

The **truncus vessel is redirected** to carry oxygenated blood to the rest of the body. The valve in the vessel is now called a "neo-aortic valve."

If there are **structural problems with the aorta** (such as interruption or coarctation), it is repaired surgically.

If there are truncal valve problems, such as leaking or narrowing, **valve repair or replacement** may be performed.

REPAIRED TRUNCUS ARTERIOSUS: LONG-TERM



***Valve Issues*:** The tube (RV-PA conduit) that connects the right ventricle to the pulmonary artery can become narrow (stenosis) or leak (insufficiency). The neo-aortic valve may develop leaking (insufficiency) or narrowing (stenosis).

Plumbing Issues:

The aorta may become enlarged (neo-aortic dilation). The pulmonary artery may develop narrowing (stenosis).

If you had a late surgical repair, there is a risk of developing increased pressure in the lungs (pulmonary hypertension).

If you had a repair for interruption of the aorta or coarctation of the aorta, there is a risk for developing recurrent narrowing of the aorta (recurrent aortic obstruction).

***Pump Issues**:* When there is RV-PA conduit or neo-aortic valve narrowing or leaking, it may enlarge (dilate) or thicken the ventricles, which may affect its pumping function.

***Electrical Issues**:* Abnormally fast heart rhythms (arrhythmias). This may be related to scarring near the surgical site.

Not everyone will experience all long-term complications or symptoms.

REPAIRED TRUNCUS ARTERIOSUS: COMMON SYMPTOMS

Possible Symptoms

Tiredness: Feeling more tired than usual.

Shortness of Breath, Chest pain: Especially during exercise.

Swelling: Extra fluid in your body or lungs.

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Why do these things happen?

Tiredness, shortness of breath, chest pain, swelling: Over time, the RV-PA conduit and the truncal valve may narrow or leak. This can affect the heart's pumping function.

Irregular heartbeats: May be a fast/slow heart rhythm, which can happen due to scarring or enlarged heart chambers.

What are the potential risks?

If the RV-PA conduit or truncal valve issues are significant and not repaired, it may lead to heart failure.

There is a risk of infection in the heart valves and heart lining (endocarditis).

Not everyone will experience all long-term complications or symptoms.

REPAIRED TRUNCUS ARTERIOSUS: MANAGEMENT

Surgical Procedures

RV-PA conduit replacement: You may require surgical replacement of the conduit.

Truncal valve repair/replacement: If there is significant valve narrowing (stenosis), you may require valve repair or replacement. *In some cases, a catheter procedure can be done.*

Significant enlargement in the aorta (aortic dilation) may need surgical repair.

Persistent, unexplained fevers and signs of heart infection: Surgical repair may be needed if symptoms persist despite treatment.

REPAIRED TRUNCUS ARTERIOSUS: MANAGEMENT

Catheter Procedures

Recurrence of significant narrowing of the aorta (previously repaired) may need a catheter procedure to enlarge it.

Pulmonary artery stenosis: To dilate (enlarge) the artery using a balloon and/or placement of a stent (mesh scaffold). *In some cases, surgery may be required.*

Arrhythmias: May need an ablation procedure. Sometimes a pacemaker or defibrillator may be necessary.

RV-PA conduit replacement: A new pulmonary valve (e.g., Melody or Harmony) may be placed within the conduit.

TRUNCUS ARTERIOSUS:
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every year

ECG/EKG, Echocardiogram:
every year

Exercise Test, Holter Monitor:
as needed

Cardiac MRI/CT:
every 3 - 5 years

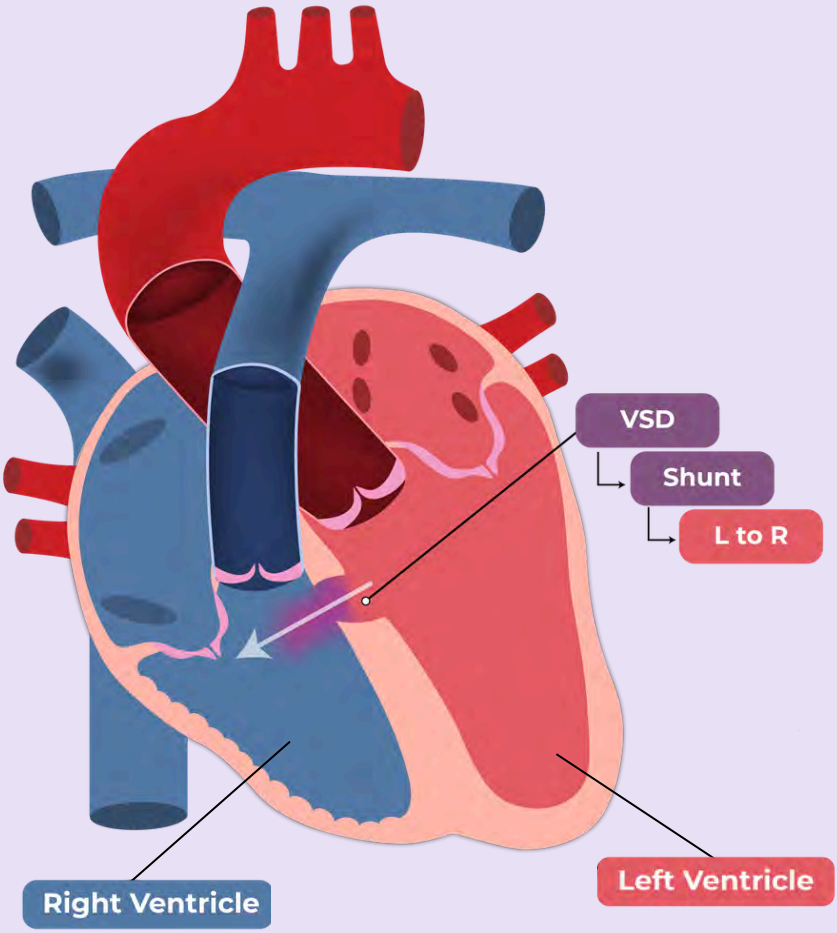
**Based on the 2025 and 2018 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs, andprovider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

WHAT IS A VENTRICULAR SEPTAL DEFECT (VSD)?

Structural

A VSD is a **hole** in the wall (septum) that separates the **two lower chambers** (right and left ventricles) of the heart.



What Happens?

Blood flows the **wrong way**. Some of the blood that should move to the body instead flows across the wall (septum) between the ventricles into the lungs and back to the left side of the heart. This is a left (L) to right (R) **shunt**.

VSD: WHAT TO KNOW

Statistics

VSDs are the most common CHD in children, representing **50% of all CHD cases** and 20 - 37% when occurring as an isolated defect.

Associated With:

Structural Issues

Multiple Combined Defects: A person may have more than one defect, such as a VSD and a coarctation (narrowing) of the aorta, or a VSD and pulmonary stenosis.

A Condition Made up of Several Defects: Some CHDs are defined by a specific set of defects that occur together. For example, Tetralogy of Fallot, Transposition of the Great Arteries, and Endocardial Cushion are some such conditions that include VSD as one of the defects.

If your VSD occurs with other heart defects, you should review the sections on those defects as well.
You will be able to receive a PDF of all the conditions at the end.

VSD:
TYPES & IMPLICATIONS

VSDs occur in different parts of the septum, and the location often determines the long-term implications and treatment considerations.

- **Membranous VSD:** located in the thinner upper part of the septum. **most common*
- **Outlet VSD** (also known as Supracristal VSD): Located in the septum near the aortic and pulmonary valves. **second most common*
- **Inlet VSD:** Located in the septum near the tricuspid and mitral valves, it is rarely isolated but is often associated with other defects, such as an atrioventricular canal defect. *Please review the AVSD section if you think you have an inlet VSD.*
- **Muscular VSD**: Located in the thick muscle of the septum. **least common*

Depending on the size of the VSD, there are differences in management and long-term implications.

VSD:
REPAIR PROCEDURES

Based on its **location, size, and complexity**, your doctor will decide whether your VSD needs to be repaired and how to repair it.

- Most VSDs close on their own by adulthood, or you may have had a procedure in childhood to close it.
- Some VSDs that don't close by adulthood may not require repair, but you still need regular follow-up care.
- If your VSD doesn't close by adulthood and your doctor determines it should be repaired, there are two possible repair options listed below.

Catheter Procedure

This less invasive procedure involves inserting a device (such as a septal occluder) through a small tube in your groin and placing it across the hole.
**Least common type of repair*

Surgical Procedure

This more invasive procedure uses a tissue patch that is surgically placed to repair the VSD.
**Most common type of repair*

***SMALL UNREPAIRED* VSD:** LONG-TERM

A small and unrepaired VSD may not cause major problems.

Different issues can occur with a small unrepaired VSD such as:

Valve Issues: A leaky aortic valve due to loss of support for the valve tissue is more common with **outlet VSD** and is sometimes also seen with **membranous VSD.** A leaky mitral or tricuspid valve due to loss of support for the valve tissue is common with an **inlet VSD.**

Structural Issues: Double-chambered right ventricle (DCRV) is a rare complication that often occurs with **membranous VSD** and is associated with increased muscle thickness in the right ventricle.

Infection Risk: There is a risk of infection in the heart valves or the septal tissues around the VSD (endocarditis).

Not everyone will experience all long-term complications or symptoms.

SMALL UNREPAIRED VSD: COMMON SYMPTOMS

You may not have any symptoms if your VSD is small and unrepaired.

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs.

Irregular hearteats: Feeling your heart racing or skipping beats (palpitations).

Why do these things happen?

If symptoms appear from a small unrepaired VSD, they are likely secondary to structural defects (severely leaky or infected valves).

Swelling: Can occur due to weak heart function, leaky valves, or high lung pressure.

Not everyone will experience all long-term complications or symptoms.

SMALL UNREPAIRED **VSD:**
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:

(outpatient)
every 3 - 5 years
(more frequent if any concern for structural issues or infection)

ECG/EKG, Echocardiogram:

every 3 - 5 years

Exercise Test:

as needed

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If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

LARGE UNREPAIRED **VSD:** LONG-TERM

Structural Issues: Because of the hole, oxygenated blood from the left ventricle flows into the lungs instead of to the body, placing strain on your heart and lungs.

Plumbing Issues: The extra blood flowing to your lungs can raise pressure in the lung arteries (pulmonary hypertension). Over time, these blood vessels become stiff, putting extra strain on the right side of the heart.

Pump Issues: The extra strain on the heart, whether left side (from extra blood) or right side (from high lung pressures), can cause the heart chambers (atria and ventricles) to become enlarged or weak.

Electrical Issues: Enlarged upper chambers (atria) can lead to abnormal heart rhythms, such as atrial flutter.

Valve Issues: An enlarged left ventricle can stretch the mitral valve, causing it to leak. In some types of VSD (such as outlet VSD), the nearby aortic or pulmonary valve may also be abnormal, leading to leakage or narrowing.

Infection Risk: There is a risk of infection of the heart valves or the septal tissues around the VSD (endocarditis).

Not everyone will experience all long-term complications or symptoms.

LARGE UNREPAIRED VSD: COMMON SYMPTOMS

You may not have any symptoms, and your VSD might be found by chance.

Possible Symptoms

Tiredness: Feeling more tired than usual.

Swelling: Extra fluid in your body or lungs.

Irregular heartbeats: Feeling your heart racing or skipping beats (palpitations).

Why do these things happen?

Tiredness: High pressure can make the blood vessels in your lungs stiff. This makes it harder for your blood to pick up oxygen, leaving you feeling exhausted even after mild activity.

Swelling: Can occur due to weak heart function, leaky valves, or high lung pressure.

Not everyone will experience all long-term complications or symptoms.

LARGE UNREPAIRED **VSD:** TREATMENT PATHS

If Lung Pressures are Normal

- The primary treatment is to repair the hole (usually requires surgery).
- You might need **medication** to get rid of **excess fluid** or to strengthen your heart.
- **Rhythm problems** can be treated with medications or, rarely, an ablation.

If Lung Pressures are High

- You may need **medications to help lower lung pressures**, such as sildenafil or macitentan.
- VSD repair is often delayed until the pressure in your lungs improves.
- You may need **blood thinners** to lower your risk of stroke.
- You might also need medications that help your heart pump more effectively.

LARGE UNREPAIRED **VSD:**
FOLLOW-UP

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 6 - 12 months

ECG/EKG, Echocardiogram:
every year

Exercise Test:
every 1 - 2 years

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

REPAIRED VSD:
FOLLOW-UP

If you were NOT TREATED for high lung pressure before repair*: Most people do well after the hole is repaired, assuming you had no other heart-related problems.*

Recommendations*

Routine Cardiologist Visits:

(outpatient)
Consider a routine outpatient ACHD cardiologist or a general cardiologist visit every ~5 years

Primary Care Visits:

Continue to follow up with your primary care doctor

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

REPAIRED VSD:
FOLLOW-UP

If you WERE TREATED for high lung pressure before repair:
You may need to continue to take some medications or slowly get off of them over time after the repair of VSD.

Recommendations*

Routine ACHD Cardiologist Visits:
(outpatient)
every 1 - 2 years

ECG/EKG, Echocardiogram, Heart Monitor:
1 - 2 years

Exercise Test:
as needed

**Based on the 2018 and 2025 ACC/AHA Guidelines for the Management of Adults With Congenital Heart Disease. Recommendations may vary based on individual needs and provider preferences.*

If you are planning for pregnancy, talk to your ACHD doctor early in your planning process.

OTHER CONDITIONS

While we have included common congenital heart defects (CHDs), there are others that are rare and usually associated with the other conditions. Here we provide a brief descriptions of these

Dextrocardia

You are born with your heart on the right side of the chest instead of the left. On its own, this is not harmful and does not need any treatment. However, some people with dextrocardia may also have other heart problems or differences in how their abdominal organs (like the stomach or liver) are positioned. If that is the case, treatment or surgery may be needed to address those additional issues.

Cor Triatriatum

You are born with an extra wall of tissue inside one of the upper chambers of the heart (atria). This extra wall can partially block the normal flow of blood, making the heart work harder than it should. Depending on how much the blood flow is blocked, some people may have no symptoms at all while others may experience breathing difficulties or other heart-related problems. If the blockage is significant, you might require a surgical procedure to remove this extra wall of tissue. The surgery is generally very effective.

OTHER CONDITIONS

Heterotaxy

Your internal organs, such as the heart, lungs, liver, and spleen, may be positioned abnormally or on the opposite side of the body from where they would normally be. Often, the organs may not have developed normally. This often occurs alongside heart defects and other problems affecting the lungs, spleen, or digestive system. Treatment depends on which organs are affected and how severely, and may range from medications to surgery to correct or manage the associated heart and organ problems.

Supravalvular Aortic or Pulmonary Stenosis

You may be born with a narrowing in the aorta just above the aortic valve (supravalvular aortic stenosis) or in the pulmonary artery just above the pulmonary valve (supravalvular pulmonary stenosis). This makes it harder for the heart to pump the blood across the narrowing. If narrowing is mild, you may have few or no symptoms. If it is severe, you may experience chest pain, shortness of breath, or dizziness. In such cases, surgery or a catheter procedure can be performed to widen or repair the narrowed area. Sometimes, these conditions may be associated with Williams syndrome or other genetic conditions.

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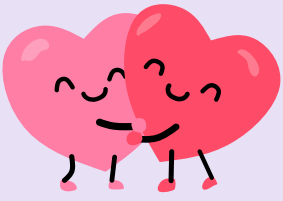

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HEART ILLUSTRATIONS

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