


# HYPOPLASTIC LEFT HEART SYNDROME

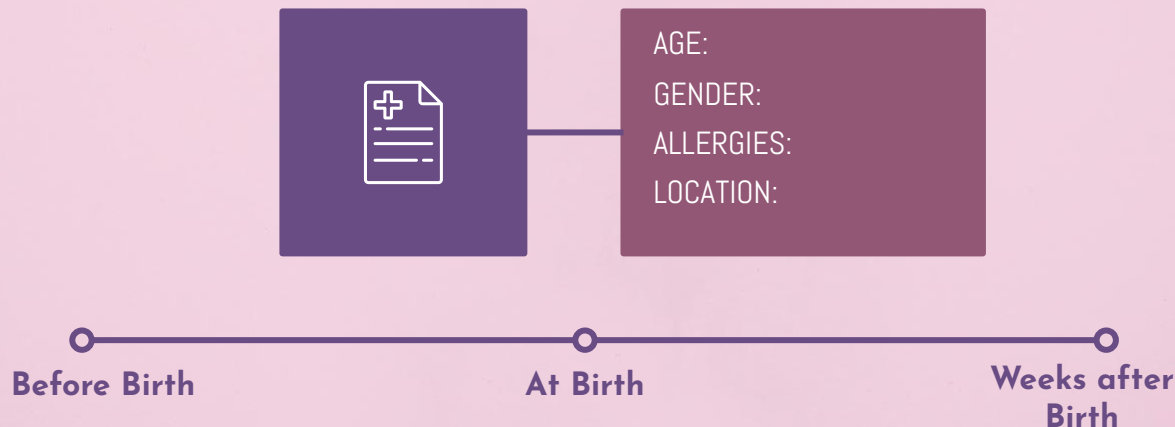


PULSE UCLA CASE STUDY

# PATIENT X MEDICAL HISTORY

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Patient X's medical information was excluded to protect patient confidentiality. This information is typically permitted to be accessible to UCLA Health volunteers and professionals only. PULSE members present this information to UCLA health volunteers and UCLA physicians only.



# WHAT IS HYPOPLASTIC LEFT HEART SYNDROME?

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## Hypoplastic Left Heart Syndrome

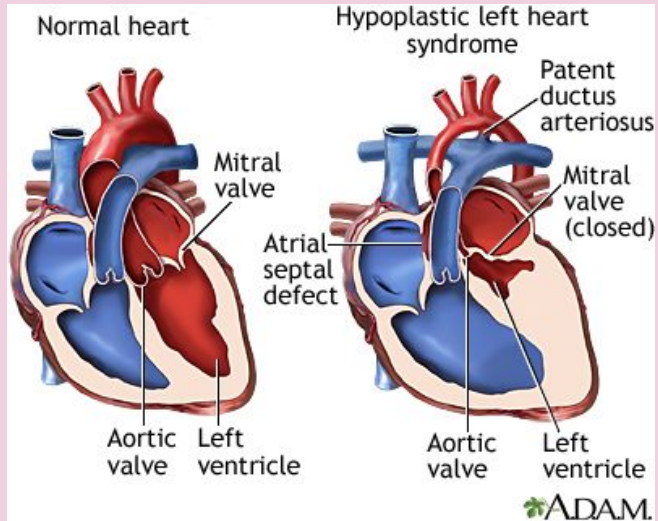
A congenital disease\* in which the left heart is extremely underdeveloped, to the point where there isn't enough pressure or room for the left side to pump blood to the body.

## Statistics

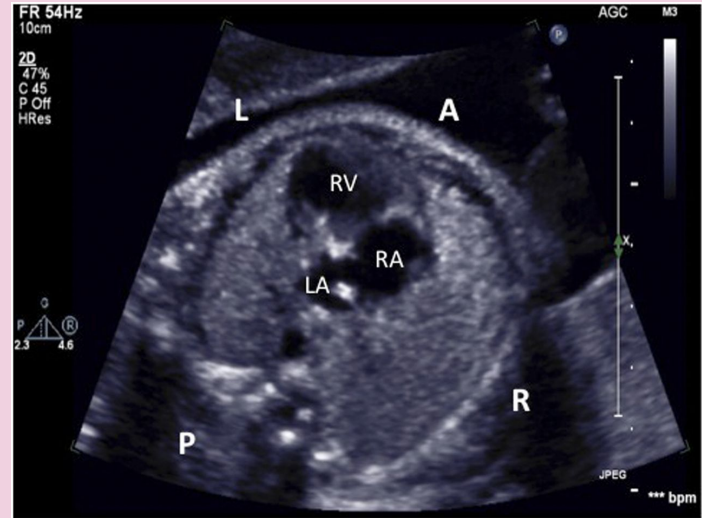
1 in 4344 babies in the U.S. are born with this syndrome every year. The cause is unknown.

**\*Congenital Disease:** Condition present at birth regardless of its cause. Birth defects may result in disabilities that may be physical, intellectual, or developmental. The disabilities can range from mild to severe.

# DIAGRAMS



Normal heart vs. Hypoplastic left heart syndrome



Ultrasound

# DIAGNOSIS

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## Before Birth

This condition can be detected through **ultrasound** before birth



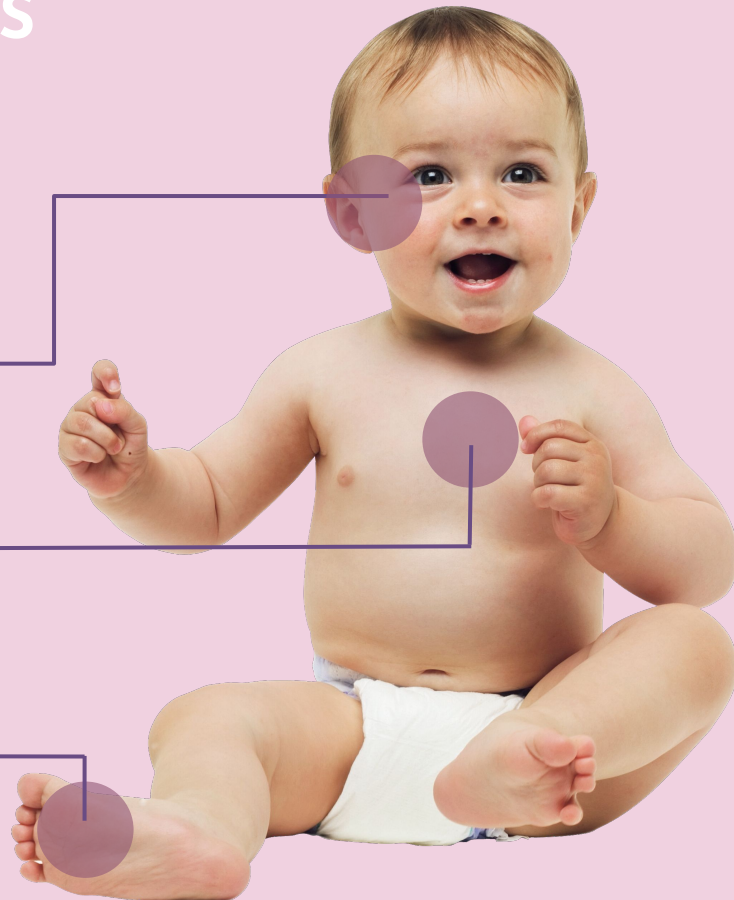
## After Birth

If the heart's condition is not found with ultrasound before birth, there are **symptoms** to serve as key indicators for identifying the condition of the baby

# SYMPTOMS

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- Drowsiness
- Rapid breathing
- Weak Pulse
- Weak Feeding
- Cyanosis (pale skin color)
- Cold hands and feet





# TREATMENT 1

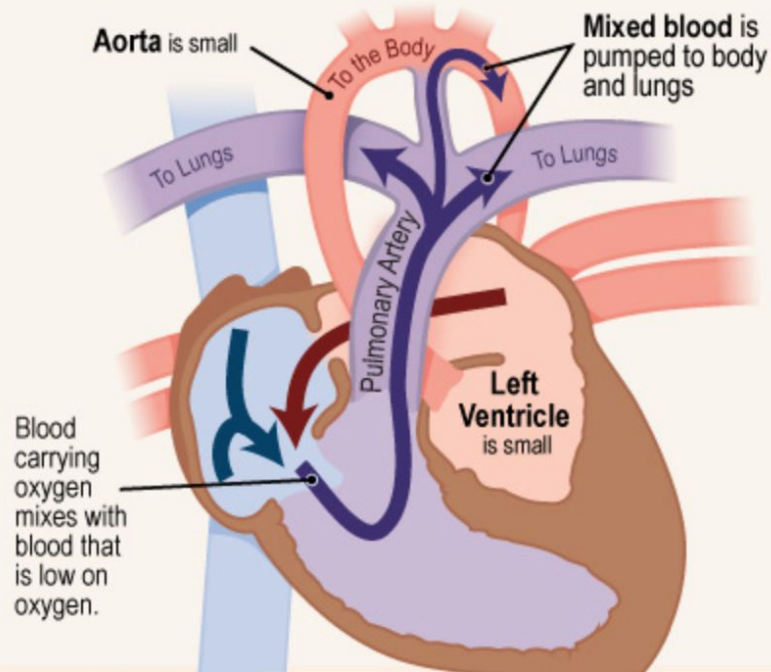
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## Norwood/Sano Surgery

1. Ideally, the baby would undergo surgery at 1-2 weeks
2. The **pulmonary artery** is cleaved and joined with the aorta
3. The **Patent Ductus Arteriosus (PDA)** connects the aorta and pulmonary artery in babies
  - a. Before surgery, PDA is preserved so deoxygenated blood can flow to the pulmonary artery followed by the lungs, which then filter the blood back into right atrium
  - b. After surgery, the PDA is removed

# NORWOOD PROCEDURE DIAGRAMS

Babies With HLHS Need Surgery  
Because of These Problems



During the Norwood Procedure

**BT shunt** is added

**Alternative**  
**Sano shunt**  
is added

**New Aorta**

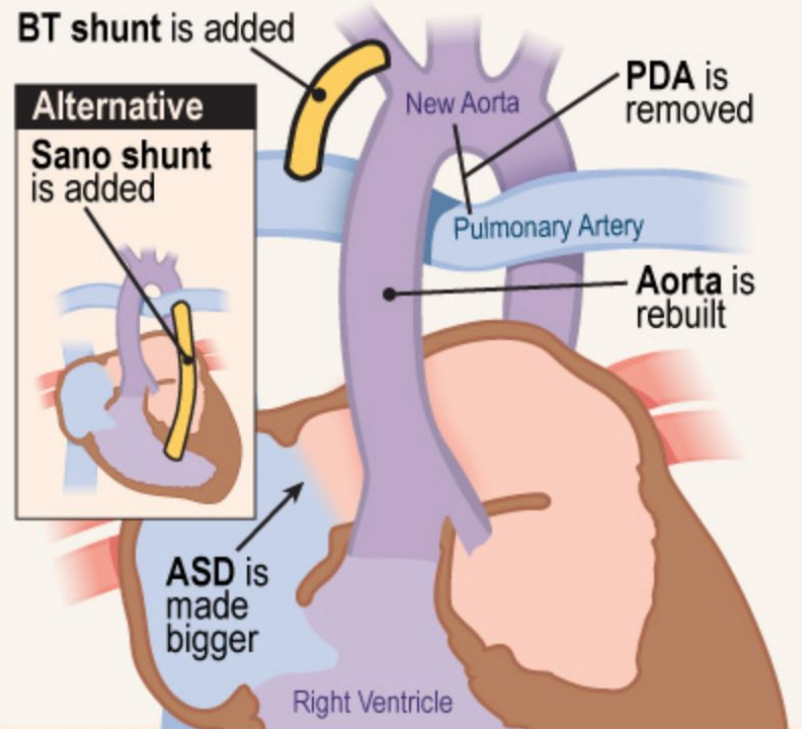
**PDA** is removed

**Pulmonary Artery**

**Aorta** is rebuilt

**ASD** is made bigger

**Right Ventricle**







## TREATMENT 2

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### Glenn Surgery

1. Ideally, the baby would undergo surgery at 4 - 6 months old
2. The **Superior Vena Cava (SVC)** is cleaved and joined with the **pulmonary artery** so that the unoxygenated blood from the upper part of the body can go straight to the lungs.
3. This allows for the right ventricle to do less work because before it was pumping both the upper body and lower body blood to the lungs.

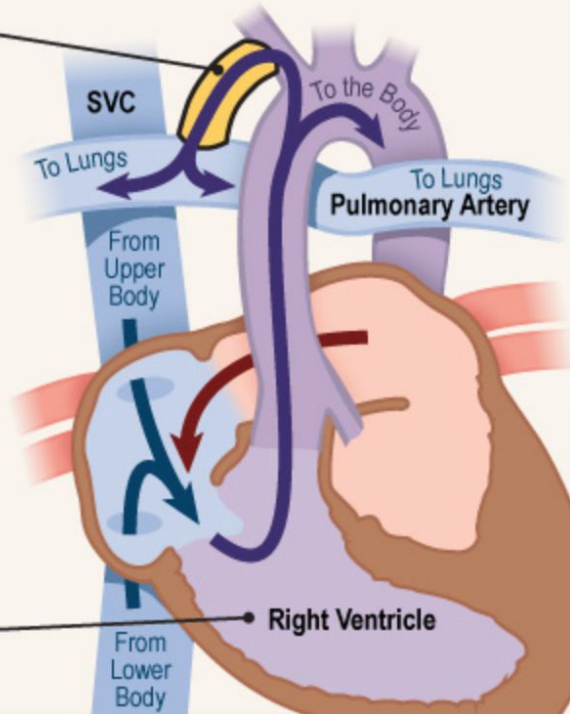
# GLENN PROCEDURE DIAGRAMS

A Child With HLHS Needs the Glenn Procedure Because

The **shunt\*** must be replaced with a more permanent solution

\*this shows one of two possible locations for a shunt

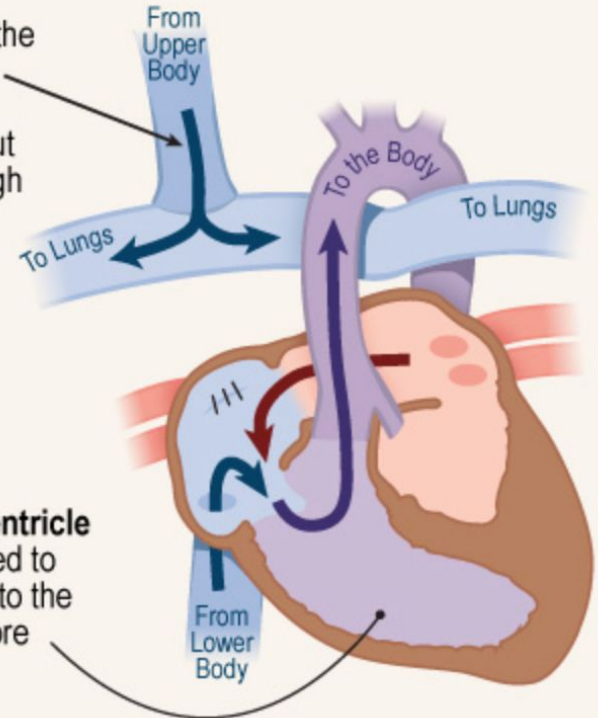
The **right ventricle** is working too hard pumping blood to the body and lungs



After the Glenn Procedure

Blood from the upper body goes to the lungs without going through the heart

The **right ventricle** does not need to pump blood to the lungs anymore





## TREATMENT 3

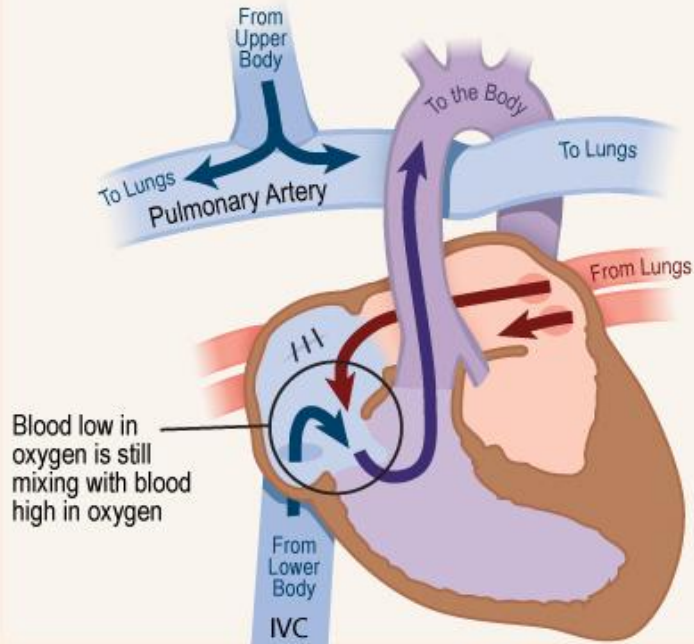
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### Fontan Surgery

1. The child would undergo surgery at 18-36 months
2. The **Inferior Vena Cava (IVC)** is disconnected from the heart and routed directly to the pulmonary artery with a conduit
  - a. Blood goes directly from lower body to the pulmonary artery and then the lungs, without having to go through the heart
3. A **Fenestration** is created between the conduit and the right atrium to allow lungs to get used to extra blood flow
4. High- and low-oxygenated blood is now separated and more oxygen can get to the body

# FONTAN PROCEDURE DIAGRAMS

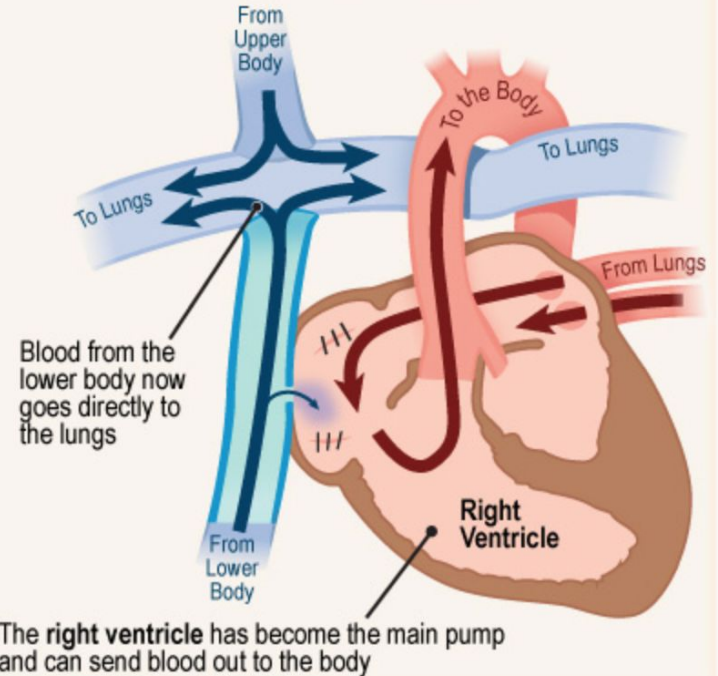
A Child With HLHS Needs the Fontan Procedure Because



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After the Fontan Procedure





# TREATMENT MEDICATIONS

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## **Furosemide**

A diuretic drug that causes one to urinate more (eliminate excess fluids)



## **Captopril**

Used to maintain a steady blood pressure



## **Prostaglandin (Before Norwood)**

Used to ensure that the PDA stays open until surgical corrections are made

# POST-NORWOOD TREATMENT

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01

## Hospital

- Around-the-clock monitoring of the patient
- Administering medications
- Teach parents how to care for baby

02

## Home

- Parents monitor baby's growth, weight gain, and oxygen levels





QUESTIONS ?

# THANK YOU

To the UCLA physicians, for allowing us to shadow in the pediatrics department.

To all the medical staff who helped us out during the quarter.

To our fellow colleagues for adding more to our experience.





# REFERENCES

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- "Hypoplastic Left Heart Syndrome." *Genetic and Rare Diseases Information Center*, U.S. Department of Health and Human Services, <https://rarediseases.info.nih.gov/diseases/6739/hypoplastic-left-heart-syndrome/cases/3158>
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