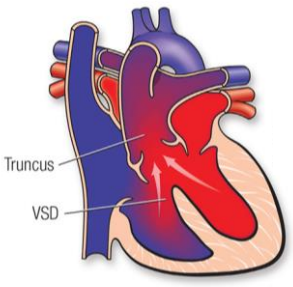
Truncus Arteriosus (TA)

Truncus Arteriosus (TA) is a congenital heart defect in which the pulmonary artery and the aorta fail to separate, resulting in a single great vessel arises from the heart, called **truncus**. In addition, there is usually a hole (**Ventricular Septal Defect, VSD**) in the septum, which normally separates the left and right lower pumping chambers (ventricles) of the heart. Please see our info sheet on Ventricular Septal Defect for more information.

- Instead of having a pulmonary valve and an aortic valve, the baby has a single valve (**truncal valve)**.
- In a baby who has truncus arteriosus and VSD, oxygen-rich blood from the left ventricle mixes with oxygen-poor blood from the right ventricle. This blood exits the heart and travels to both the lungs and the rest of the body. As a result, the lung has too much blood, and the heart has to work hard to pump blood to the rest of the body.

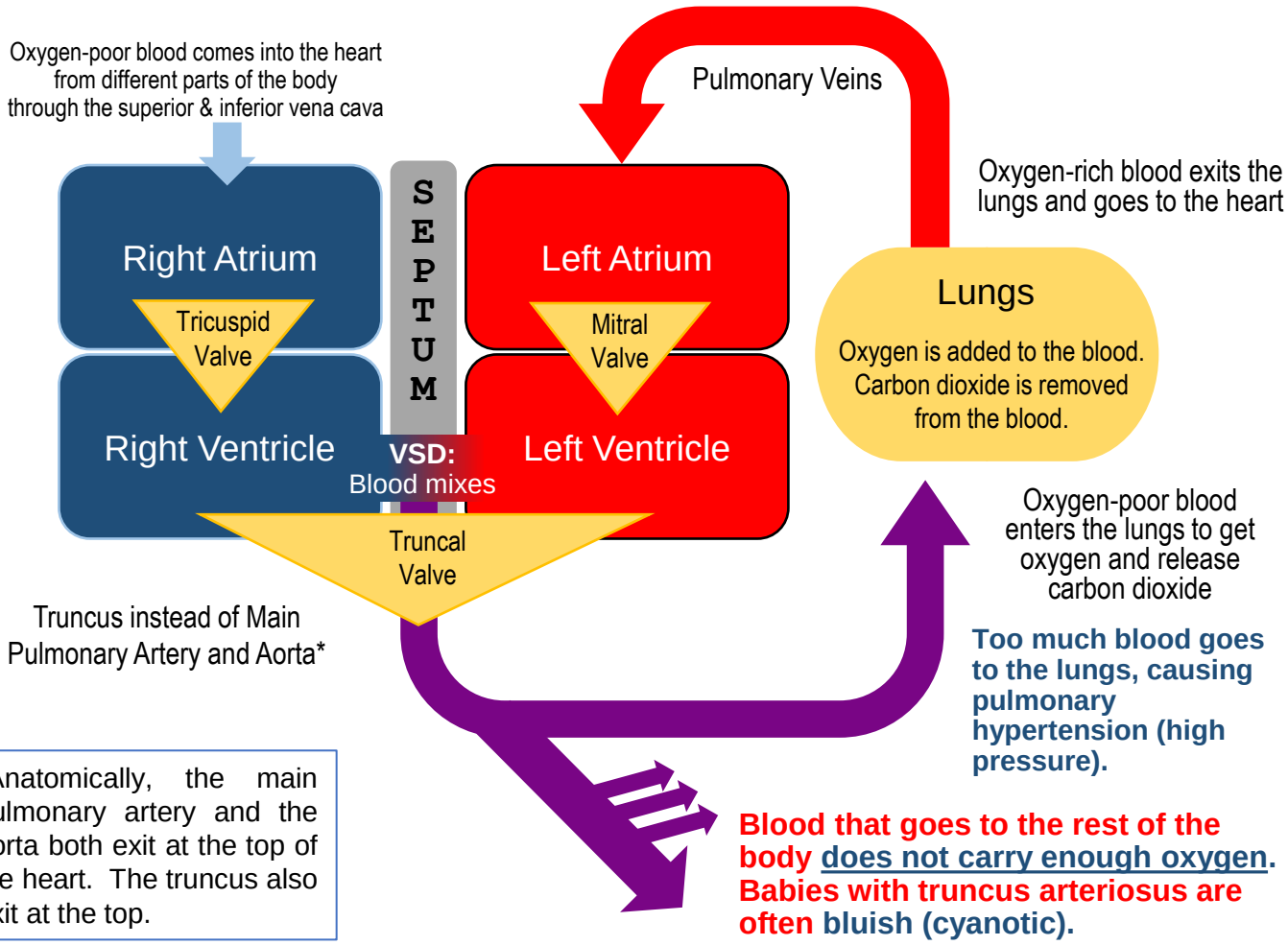


Please click on the image to see a larger version on the website of the American Heart Association

A Schematic Diagram of the Heart with Truncus Arteriosus

Shown as if you are facing the patient; Not to scale

Please also see our schematic diagram of The Heart and Normal Blood Flow for comparison.



*Anatomically, the main pulmonary artery and the aorta both exit at the top of the heart. The truncus also exit at the top.

Truncus Arteriosus (continued)

Truncus Arteriosus (TA) and Individuals with 22q Differences

- Of all patients with TA, 30 to 50% have 22q11.2 deletion syndrome (22q11.2DS).
- 5 to 10% of children with 22q11.2DS are born with TA.
  - In patients with 22q11.2DS, TA can happen together with other heart defects.
- In a study, 2 out of 85 children with 22q11.2 duplication syndrome (22q11.2DupS) were born with TA.

Diagnosis for Truncus Arteriosus

- During pregnancy, TA can be diagnosed using **ultrasound** and **fetal echocardiogram**.
- After birth, TA can be diagnosed using **echocardiogram**. Other examinations such as MRI or CT may be useful for diagnostic confirmation and preoperative management.
- Babies with TA are often in **respiratory distress** and can look **bluish**. They can also have symptoms such as difficulty breathing, increased heart rate, weak pulse, poor feeding, and extreme sleepiness.

Treatment for Truncus Arteriosus

- Some children may need medications (e.g. diuretics to reduce the pulmonary overflow)
- Babies who are not gaining weight may need a high-calorie formula or even a feeding tube.
- **Surgery** is necessary to close the hole and separate the vessels.
  - The ventricular septal defect is closed with a patch in a way that places the TA to the left of the septum, where it functions as the aorta only.
  - The pulmonary arteries are disconnected from the truncus and connected to a valved conduit from the right ventricle.

Ongoing care for Truncus Arteriosus

1. As time goes on, the child may outgrow the conduit, or the conduit may become blocked. The aortic valve (the old truncus valve) may also become leaky. Additional surgeries may be needed.
2. **Follow-up with a cardiologist**. He/she may recommend **tests** for ongoing monitoring.
3. Some people may need medications.
4. Even after the repair, some individuals **may need to avoid intense sports**. Please discuss exercise tolerance with your cardiologist.
5. Some people may need **endocarditis prophylaxis** – This means taking antibiotics before certain dental procedures to prevent bacteria from causing infections in the heart. Please discuss with your cardiologist.
6. Women who have repaired TA and are considering **getting pregnant should discuss the risks** with their healthcare providers. Those with problems with the valves and pulmonary hypertension are at a much higher risk. Please discuss with your cardiologist

For more info, please visit the websites of the CDC or the American Heart Association.

22q International 22q11.2 Foundation

The mission of the International 22q11.2 Foundation is to improve the quality of life for individuals affected by chromosome 22q11.2 differences through family and professional partnerships.
This information is brought to you by the Foundation for educational purposes only. It is not intended to be taken as medical advice. If you have concerns, please talk to your healthcare provider.

Joanne Loo, PhD & Carolina Putotto, MD, 02/2022