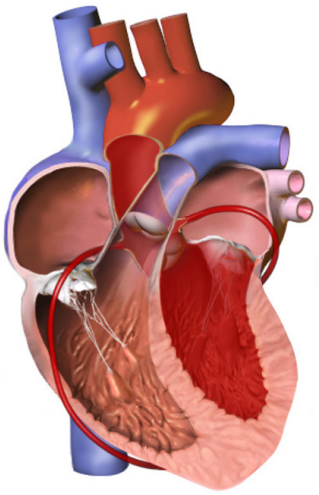

NATIONAL CENTER FOR CASE STUDY TEACHING IN SCIENCE

Cardiac Conundrum:

Cardiac Anatomy, Fetal Circulation, and Congenital Heart Disease

by

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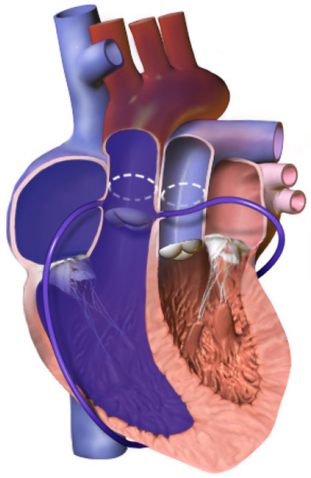

Pre-Class Preparation

If you are unfamiliar with human cardiac anatomy and the flow of blood through the heart, please watch the video listed below.

- Neural Academy. (2018). Blood flow through the heart in two minutes. <https://youtu.be/jBt5jZSWhMI>

Part I – The First Look

Callie and Nate walked arm and arm through the doors of the obstetrics building and into the waiting room (obstetricians are doctors who specialize in pregnancy). The couple had excitedly awaited their initial ultrasound appointment since discovering that they were pregnant with their first child. They eagerly anticipated seeing the black and white image of the first trimester embryo.

After leafing through magazines in the waiting room, Callie and Nate were called back by Jenna, an ultrasound technician, and followed her into a patient room. Callie took a seat on the exam bed as Jenna greeted them, asking "How are you two feeling about the pregnancy?"

"So excited!" Callie replied. "We can't wait to see the image of our little baby on the screen."

Nate helped Callie lay back on the bed and asked, "Will we be able to hear the heartbeat at this ultrasound? When my sister in-law, Linda, was pregnant, my brother said it was the most amazing thing he had ever heard."

Jenna glanced down at their paperwork before replying. "Based on the estimated time from your last menstrual period, it looks like this is an eight-week ultrasound. I can't make any promises, but the embryo's heartbeat can usually be seen and heard starting at about five to six weeks."

Callie smiled at Nate as Jenna applied a cool gel on her belly and began the ultrasound. Jenna measured "crown to rump" length to establish the baby's due date and soon after, the expecting parents were listening to the rhythmic lub-dub of their baby's heartbeat.

"That's amazing!" Callie exclaimed.

"My heart feels like it's jumping for joy!" echoed Nate.

Jenna smiled at the couple's excitement over the heartbeat of their little embryo and printed out some copies of the ultrasound for them to take home. Callie and Nate thanked Jenna for the sonograms and grinned ear to ear as they walked out to the car.

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Questions

1. Match each major structure of the heart to its corresponding letter in Figure 1. Write the corresponding letter and oxygenation status to the left of the word. Use the abbreviation "OxR" for a structure containing oxygen-rich blood and "OxP" for a structure containing oxygen-poor blood. "N/A" refers to structures that do not carry blood (i.e., are not chambers nor vessels that are filled with blood). The left atrium has been completed as an example.

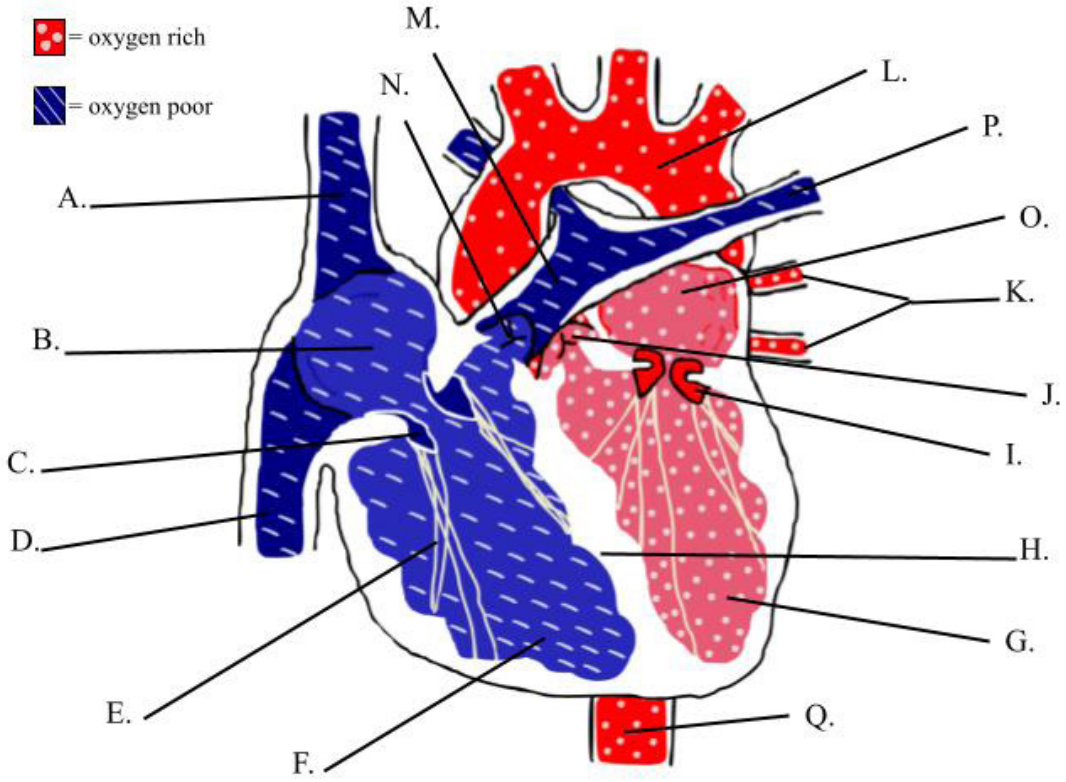

Figure 1. Anatomy of the human heart. *Credit:* Julia G. Primak.

Left Atrium **O, OxR**

Left Ventricle

Right Atrium

Right Ventricle

Superior Vena Cava

Inferior Vena Cava

Left Pulmonary Artery

Aorta

Chordae Tendineae **N/A**

Bicuspid Valve

Tricuspid Valve

Pulmonary Semilunar Valve

Pulmonary Veins

Aortic Semilunar Valve

Interventricular Septum **N/A**

Pulmonary Trunk

Descending Aorta

2. To supply oxygen-rich blood to the upper portion of the body, from right to left, the aorta branches into the brachiocephalic, left common carotid, and left subclavian arteries. The brachiocephalic artery then branches further into the right common carotid and right subclavian arteries. The aortic arch is continuous with the descending aorta, which supplies the lower portion of the body with oxygen-rich blood. Draw a diagram of the aorta that includes these vessels:

   - Brachiocephalic Artery
   - Left Subclavian Artery
   - Descending Aorta
   - Right Common Carotid Artery
   - Left Common Carotid Artery
   - Right Subclavian Artery

3. Use the following terms to describe the flow of oxygen-poor and oxygen-rich blood through the heart:

   - Aorta
   - Aortic Semilunar Valve
   - Inferior Vena Cava
   - Superior Vena Cava
   - Right Atrium
   - Right Ventricle
   - Left Atrium
   - Left Ventricle
   - Mitral/Bicuspid Valve
   - Tricuspid Valve
   - Pulmonary Semilunar Valve
   - Pulmonary Arteries
   - Pulmonary Veins

Oxygen-poor blood enters the _____ _____ through the _____ _____ _____ and _____ _____ _____. The blood then goes through the _____ _____ and enters the _____ _____. This blood is then pumped through the _____ _____ _____ and travels through the _____ _____ into the lungs.

Oxygen-rich blood returns to the heart from the lungs through the _____ _____ and enters the _____ _____. Blood travels through the _____ _____ into the _____ _____. The blood is then pumped through the _____ _____ _____ and into the _____ where it then circulates to the rest of the body.

Part II – Fetal Flow

Shortly before their four-month ultrasound, Nate and Callie arrived home from the grocery store on a sunny Saturday afternoon to the sight of a package sitting on their front porch.

"I wonder if that is the book about pregnancy your sister-in-law has been talking about," said Callie.

The couple brought the box inside and opened it up. Sure enough, there was the book.

Callie read the title aloud: "What to Envision When You're Eating for Two: The Ultimate Pregnancy Guide."

Nate laughed at the title. He didn't understand why Linda insisted he buy it for Callie. Couldn't they just use the internet? But, alas, books were helpful too.

Callie sat down on the couch and began to read through the book with Nate. It wasn't long before they reached the chapter on fetal circulation.

"I still can't believe we got to hear the baby's heartbeat at the eight-week ultrasound," Callie remarked.

As Callie and Nate continue to read, they realized that the way blood circulates through their baby was very different from the way blood moved through their bodies. First they read that the structure of a fetus' heart differs from the structure of their adult hearts. Then they learned that the fetus "breathes" through the placenta, not its developing lungs. They decided to call Callie's younger brother, Michael, a nursing student currently taking an anatomy class. Callie and Nate asked Michael if he could help them understand the heart structure and circulation of their baby.

"I'm so glad you guys called!" said Michael. "We actually just learned about fetal cardiac anatomy and circulation, and I've been thinking about my little niece or nephew this whole time."

Questions

Watch the following video to learn about fetal blood circulation. Then answer the questions below to help Michael teach Callie and Nate about the anatomy of their baby's heart and how fetal blood circulation works.

- Byte Size Med (2020). Foetal (fetal) circulation before and at birth: cardiac physiology, embryology. Running time: 8:08 min. <https://youtu.be/m3p5PsB6aZ4>

4. Summarize fetal blood flow by creating a verbal flow chart or drawing that depicts the flow of blood from the placenta to the fetus and back to the placenta. Label where the blood is oxygen rich, oxygen poor, and oxygen mixed (when there is both oxygen-rich and oxygen-poor blood). Be sure to include all the following structures:

• Aorta	• Liver	• Right Ventricle
• Ductus Arteriosus	• Placenta	• Systemic Tissues
• Ductus Venosus	• Pulmonary Trunk	• Umbilical Arteries
• Foramen Ovale	• Right Atrium	• Umbilical Vein

5. Complete the following table to compare blood flow in the fetus to blood flow in an adult. For the purpose of this table, ignore any valves.

Table 1. Comparison of blood flow in fetus and adult.

Point of Comparison	*In the Fetus*	*In the Adult*
Blood from the right atrium moves to the . . .		
Blood from the right ventricle moves to the . . .		
Blood from the left atrium moves to the . . .		
Blood from the left ventricle moves to the . . .		
Blood from the pulmonary arteries moves to the . . .		
Blood loads oxygen and unloads carbon dioxide in the…		

6. Where is the foramen ovale located?
   a. Between the right and left atrium.
   b. Between the right and left ventricles.
   c. In the interventricular septum.
   d. In the aorta.

7. Which of the following is the function of the foramen ovale?
   a. To allow oxygen-rich blood to pass from the left ventricle to the right ventricle.
   b. To allow oxygen-mixed blood to pass from the right atrium to the left atrium.
   c. To allow oxygen-rich blood to pass from the right ventricle to the aorta.
   d. To allow oxygen-mixed blood to pass from the left ventricle to the left atrium.

8. Which of the following is the function of the ductus arteriosus?
   a. To allow oxygen-mixed blood to flow from the aorta into the pulmonary arteries.
   b. To allow oxygen-mixed blood to flow from the pulmonary arteries into the aorta.
   c. To allow oxygen-rich blood to flow from the umbilical vein to the liver.
   d. To allow oxygen-rich blood to flow from the liver to the right atrium.
   e. To allow oxygen-mixed blood to flow into the lungs to be oxygenated.

9. Through which of the following structures does blood enter the right atrium of the fetal heart? Circle all that apply.
   a. Ductus Arteriosus
   b. Ductus Venosus
   c. Inferior Vena Cava
   d. Superior Vena Cava

10. Circle the correct answers in the following paragraph:

    In fetal circulation, oxygen poor blood from the systemic tissues travels through the umbilical _____ (arteries/veins) to the placenta. Gas exchange occurs in the placenta. The oxygen- _____ (rich/poor) blood then travels through the umbilical _____ (artery/vein) back to the fetus. The umbilical vein then branches into the portal sinus, which brings blood to the liver. However, the _____ _____(ductus arteriosus/ductus venosus/foramen ovale) causes the majority of blood to bypasses the liver, to bring the oxygen-rich blood into the _____(right/left) _____(atrium/ventricle). Blood from the right atrium travels to both the _____ _____ (right atrium, right ventricle, left atrium, left ventricle) and the _____ _____(right atrium, right ventricle, left atrium, left ventricle). The _____ _____ (ductus arteriosus/ductus venosus/foramen ovale) is the hole that allows blood to pass from the right atrium to the left atrium. Blood from the left atrium travels the left _____(atrium/ventricle) to be pumped into the aorta. The blood from the right ventricle is pumped into the pulmonary _____ (arteries/veins) just like adult circulation. However, since the lungs are full of amniotic fluid, the resulting pressure gradient causes the blood to pass through the _____ _____(ductus arteriosus/ductus venosus/foramen ovale) into the aorta. The aorta pumps the oxygen-rich blood to the systemic tissues and the cycle repeats.

11. Write Michael's response to Callie and Nate explaining how adult and fetal circulation differ from one another. Use your answers to the previous questions to guide you.

Part III – Family Ties

Callie and Nate returned to the obstetrician's for Callie's 23-week ultrasound. Neither of them could believe how fast the pregnancy was going and were trying to soak up every moment.

"Did you know that our baby is now the size of a sweet potato?" Nate asked as he smiled and waved a pamphlet relating the size of a fetus to different fruits and vegetables.

Callie laughed and rubbed her growing belly. "Well, our little one can kick pretty hard for a potato."

Nate glanced at the pamphlet she had been intently focused on.

"'Congenital Heart Defects.' Now that's a lively title," he remarked.

Callie sighed. "Well, I want to be as informed as possible. You never know what could end up being important. We can never be too prepared."

Nate's smile faded. "You know what, now that you mention it, I feel like I remember my mom mentioning something about her dad's brother having some kind of strange heart disease. He passed away long before I was born, but I wonder if it could be genetic?"

The expecting parents looked at each other with concern.

Nate continued, "I think his heart condition may have had something to do with the word 'great.' I don't remember exactly what it was though."

Callie looked back down at the pamphlet and traced her finger down the page. "Could it have been this? Transposition of the great arteries?"

"That sounds really familiar. What does it say?" asked Nate.

"It says that transposition of the great arteries (TGA) occurs when the outflow vessels of the heart, the pulmonary trunk and the aorta, are associated with the wrong ventricle. Essentially, the pulmonary trunk and aorta swap, flowing out of the incorrect ventricle. It doesn't harm the baby while they are in the womb, but it can cause significant symptoms after delivery. Look, here's a picture." Callie pointed to a figure in the pamphlet (see Figure 2, right).

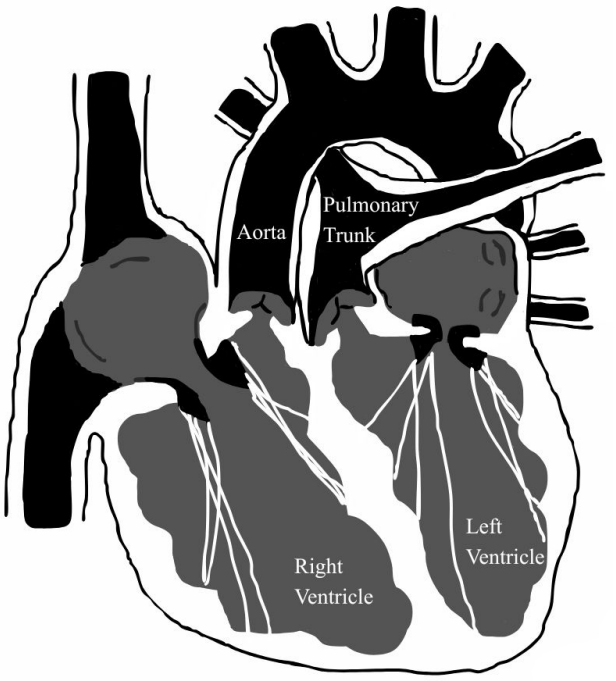

Figure 2. Anatomy of a heart with transposition of the great arteries. *Credit:* Julia G. Primak.

Question

12. Use your knowledge of fetal blood circulation to theorize as to why transposition of the great arteries is not a problem in the womb but is fatal outside of the womb.

Part IV – A Daunting Diagnosis

Nate helped Callie get comfortable on the exam table for her ultrasound as her obstetrician, Dr. Singh, flipped open her chart and offered a warm greeting. "How are you feeling today Callie? Excited to be 23 weeks already?"

"I'm doing pretty well," Callie replied. "My morning sickness has gotten much better, and I can't believe I'm already over half way done. I do have one concern that I'd like to discuss though."

"I'm all ears," said Dr. Singh.

"We were just reading the pregnancy pamphlet in the waiting room about congenital heart defects," explained Callie. "My husband thinks that his grandpa had a baby brother who passed away soon after he was born from a condition called transposition of the great arteries. Do you think our baby might be affected by this?"

"Thank you for bringing this concern to my attention. It isn't fully known if there is a genetic component to transposition of the great arteries, but since there is a family history of the condition, I would like to run an additional test that will allow us to take a better look at the structure of your baby's heart," replied Dr. Singh.

"What kind of test?" asked Nate.

"Transposition of the great arteries, TGA, is not typically detected when performing a routine ultrasound. Instead, we can use a fetal echocardiogram to allow us to look at the health of the baby's heart in much greater detail," she explained. "I'll put in an order before you guys leave today, and you should be able to schedule an appointment for one in the next week or two."

A week later Callie and Nate arrived at their fetal echocardiography (also known as an ECHO) appointment filled with anxiety. What if their baby had a heart defect?

For such a significant test, Callie was surprised when the process was over after only 30 minutes. A short time later, Dr. Singh came into the room with the results. "Callie, Nate, based on the results from the fetal echocardiogram, I'm sorry to tell you that your baby does have transposition of the great arteries."

Nate wrapped his arms around Callie as they tried to absorb the news.

"Thankfully," Dr. Singh continued, "since you brought this to our attention and we were able to diagnose your baby before birth, we can begin preparing for treatment right away. When transposition of the great arteries is diagnosed in utero, we can be ready to take immediate action to correct the defect and restore normal anatomy right after birth."

Dr. Singh informed Callie and Nate that their baby would need corrective surgery shortly after birth. The physician provided them with the information of a neonatal surgeon, Dr. Mohler, who would join their care team.

Questions

13. How would you react to the diagnosis Nate and Callie received from Dr. Singh?

14. How would you help expecting parents deal with the fact that their child will need immediate surgery after birth?

15. Put yourself in the shoes of the neonatal surgeon. What could you do to restore normal blood flow for Callie and Nate's baby?

Part V – Cardiac Correction

On a cloudy afternoon at 39 weeks pregnant, Callie waddled out to the car with Nate. Dr. Singh and Dr. Mohler had recommended that Callie schedule an induction (medically induced labor) so they could make sure they had all the proper equipment and personnel in place when the baby was born. As Callie got settled into the front seat, Nate ran back and forth to the house grabbing the bags they packed for the hospital.

"I don't think I'll need seven pillows, Nate," Callie called out.

Nate laughed as he closed the trunk and got in the car. "I just want you to be comfortable. I can't believe the time is finally here!"

"Hold your horses, hotshot," cautioned Callie. "The next few days are not going to be the easiest of our life."

Nate knew Callie was referring to more than just labor and delivery. At a previous appointment, Dr. Mohler had walked them through what to expect. Immediately following delivery, their baby would have imaging tests to confirm the diagnosis and look for additional structural defects. Assuming their baby did have transposition of the great arteries (TGA), the doctors would then perform a procedure called a balloon atrial septostomy. The procedure used a tiny balloon to reopen their baby's foramen ovale so that oxygen-rich and oxygen-poor blood could mix. Then, after their baby was stable, a corrective surgery called an arterial switch operation (ASO) would be performed. Callie was right, the next couple days might not be easy, but it would all be worth it when they could bring their baby home.

That afternoon Callie and Nate's baby was finally born.

"It's a boy!" exclaimed Dr. Singh.

Nate held Callie's hand tightly, tears of joy and worry streaming down both of the new parents' faces. They could see a faint blue tint on their baby boy's fingers as Dr. Singh handed him off to Dr. Mohler and her team to perform the necessary diagnostic imaging tests. As Callie and Nate were being moved to a recovery room, the diagnosis of TGA was confirmed. Dr. Mohler administered prostaglandin E1 to keep the baby's ductus arteriosus open. She and her team then began the balloon atrial septostomy procedure.

Two days after Callie and Nate's baby boy was born, Dr. Mohler decided the baby was stable enough for surgery. She performed the delicate arterial switch operation to correct the tiny baby's heart anatomy and save his life.

Questions

Watch the following video that showcases the most common procedure used to correct transposition of the great arteries, an arterial switch operation. Then answer the following questions about this type of surgery and the treatment Callie and Nate's baby received after he was born.

- Children's Heart Surgery Fund (2022). Transposition of the great arteries. Running time: 4:13 min. YouTube. <https://youtu.be/v5nvkmu1PYA>

16. When Callie and Nate's baby was born, his skin had a blue tint. This is a classic symptom of TGA. Circle the following word that correctly describes this appearance:

    Jaundice Cyanosis Erythema

17. Before performing a corrective surgery, Dr. Mohler gave the baby prostaglandin E1 and did a balloon atrial septostomy. What is the goal of each of these procedures, and how do they help a baby born with TGA? (If you need support answering this question, it is suggested you rewatch the video from Part II, focusing on changes after birth.)

18. In an arterial switch operation, where is the pulmonary trunk being removed from? Where is it being attached to?
    a. Removed from the left ventricle; attached to the right ventricle.
    b. Removed from the right ventricle; attached to the left ventricle.
    c. Removed from the left atrium; attached to the right atrium.
    d. Removed from the right atrium; attached to the left atrium.

19. In an arterial switch operation, where is the aorta being removed from? Where is it being attached to?
    a. Removed from the left ventricle; attached to the right ventricle.
    b. Removed from the right ventricle; attached to the left ventricle.
    c. Removed from the left atrium; attached to the right atrium.
    d. Removed from the right atrium; attached to the left atrium.

20. It is recommended to perform the arterial switch operation within two weeks of birth of a baby born with TGA. Waiting longer before operation will result in left ventricular atrophy (reduced size and number of myocardial cells due to cell death). Why might this occur?

Part VI – A Sigh of Relief

Following his successful arterial switch operation performed by Dr. Mohler, Callie and Nate visited their baby boy in the neonatal intensive care unit (NICU). They were overcome with happiness at being able to hold their baby's tiny hands and were relieved to no longer see a blue tint in his skin.

Callie beamed. "He's so perfect."

As the proud new parents sat with their baby, one of the NICU nurses came by to check on them. She asked if they had decided on a name for their new bundle of joy.

"We sure have," said Nate. "His name is Caleb."

Callie smiled, not taking her eyes off of Caleb. "The name Caleb means 'whole heart.'"

Thanks to the surgery and doctors, Caleb's whole heart was now functioning.

Two weeks later, after Caleb passed all of his NICU tests, Callie, Nate, and Caleb met with Dr. Mohler to discuss what a future with TGA after corrective surgery would look like.

"I am happy to say that the arterial switch operation Caleb received was successful," Dr. Mohler informed the new parents. "This surgery has a very low reoperation rate with a survival rate over 95% at 25 years post operation. Caleb will likely be able to lead a very normal life."

Callie and Nate both breathed a sigh of relief and smiled down at their baby boy as Dr. Mohler continued. "A few things to keep in mind are that Caleb may have reduced exercise capacity and a higher chance of being diagnosed with attention deficit/hyperactivity disorder (ADHD). However, his activities of daily living should not be noticeably impacted, and there will be many services to support his health. The last thing to note is that long-term follow-up appointments will be critical to assess heart function and detect any valve regurgitation."

Callie and Nate thanked Dr. Mohler again for all her help, and the surgeon wished them well as they made their way out of the hospital with Caleb.

Questions

21. Dr. Mohler cautioned Callie and Nate that one of the reasons Caleb would need regular follow up is to detect possible valve regurgitation (a condition where a heart valve does not close properly). Mitral valve prolapse is a specific type of valve disorder where the mitral (bicuspid) valve does not close properly. It affects 2–3% of the general population. Depending on the severity of regurgitation, mitral valve prolapse can lead to serious complications including sudden cardiac arrest. Based on your knowledge of heart structure and blood flow, how would this condition affect heart function? What are some implications of this condition?

Part VII – Real World Applications

To learn more about individuals who have had transposition of the great arteries, read the following real-life stories of a boy named "Max" and a girl named "Sophia" who were both diagnosed with TGA.

- Transposition of the great arteries: Max's story. (2009). [Webpage]. Children's Hospital of Philadelphia. <https://www.chop.edu/stories/transposition-great-arteries-maxs-story>
- Sophia's story of CHD. (n.d.). [Webpage]. Victor Chang Cardiac Research Institute. <https://www.victorchang.edu.au/patients/sophie>

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