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SINGLE VENTRICLE PHYSIOLOGY EPISODE 1: TRICUSPID ATRESIA

Developed by DR. SABINE LAGUË and DR. SHREYA MOODLEY for PedsCases.com.
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Introduction:

Hello, my name is Sabine Laguë and I am 1st year pediatrics resident at the University of British Columbia in Vancouver, Canada. I am joined on this podcast today by Dr. Shreya Moodley, a pediatric cardiologist at BC Children's Hospital in Vancouver. I would like to thank Dr. Moodley for her guidance in putting together this podcast, as well as PedsCases.com for their constructive feedback on the script and for this exciting avenue to discuss this interesting topic.

This is the first of three episodes in a series on single ventricle physiology. We are very excited about this series because now more than any time in history we are seeing an increasingly growing number of individuals living and thriving into adulthood with single ventricle physiology post-Fontan palliation. Regardless of the type of medicine you are currently or will end up practicing, it is possible that you will end up working with individuals from this population. Thus, an awareness of Fontan physiology and the systemic complications of this circulation is important.

This first episode focuses on tricuspid atresia, a relatively common form of single ventricle anatomy. The second episode discusses hypoplastic left heart syndrome, another common form of single ventricle anatomy. Both episodes follow the patient from birth through to childhood. The third episode revisits both of these patients in their late adolescence and discusses long-term management and complications of patients post-Fontan.

Please see the script available on the Pedscases website for helpful figures and diagrams to help you better understand the underlying cardiac physiology. These are also referenced in the show notes.

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The objectives of this podcast are to:

1. Describe tricuspid atresia and its anatomy-associated anomalies and pathophysiology.
2. Discuss common presenting symptoms and physical exam findings of a patient with tricuspid atresia.
3. Compare modalities used to diagnose and monitor tricuspid atresia both antenatally and postnatally.
4. Outline initial medical management of a neonate with tricuspid atresia.
5. Discuss the three stages of surgical management to ultimately achieve the Fontan circulation

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You are a 3rd year medical student at a maternal fetal medicine clinic as part of your obstetrics and gynecology clerkship rotation. You learn that the next patient you will be seeing is Sarah, a G1P0 woman at 30 weeks gestation with a singleton pregnancy. Sarah is followed because a routine detailed anatomical ultrasound that was completed at 20 weeks identified tricuspid atresia in her fetus. She was referred to a pediatric cardiologist who specializes in fetal echocardiography and works closely with Maternal Fetal Medicine. Your preceptor recommends that you go and read up on single ventricles and on tricuspid atresia, as you have not yet completed your pediatric core rotation and the fetal cardiologist attending the appointment today loves asking students questions.

You read that congenital heart disease is relatively common, occurring in 1% of the general population, or 8-12 of every 1000 live births.[1] Anatomically single ventricles comprise <2% of congenital heart defects,[2] while functionally single ventricles, such as tricuspid atresia make up 1.5-3%.[3] Males are affected by single ventricle pathology more than females, comprising 55-67% of diagnoses.[4,5]

In practical terms, single ventricle physiology is a term attributable to any circulation in which there is only one functional pumping chamber. There are a variety of anatomical variations, such as tricuspid atresia or hypoplastic left heart syndrome.

You go on to read specifically about tricuspid atresia, and then enter the room where the maternal fetal medicine specialist and the fetal cardiologist are reviewing Sarah's most recent ultrasound. The fetal cardiologist extends her hand in greeting.

Preceptor: Welcome! I'm glad that you will be sitting in on reviewing this interesting case. I hear that you've had a chance to do some reading on tricuspid atresia. Can you walk me through the basics this lesion?

Student: Tricuspid atresia is an agenesis or rudimentary development of the tricuspid valve **(Figure 1)**. Ultimately this means that there is no direct connection between the right atrium and right ventricle, and that there is usually a fibromuscular or muscular diaphragm instead of a valve.[6] This can lead to a degree of right ventricular hypoplasia.

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Preceptor: Correct. Tricuspid atresia is the third most common cyanotic heart defect following tetralogy of Fallot and transposition of the great arteries. It's also almost always associated with other cardiac anomalies. Do you know which ones?

Student: I was reading that tricuspid atresia is always associated with an atrial septal defect or a patent foramen ovale, known as an obligatory shunt, which is required for survival because it allows for blood flow to exit the right atrium when the tricuspid valve does not open. It's also associated with a ventricular septal defect, which allows for communication between the right and left ventricles. Pulmonary stenosis or atresia, which ultimately determines the amount of pulmonary blood flow, can also be seen.[6] Other more complex forms of tricuspid atresia also exist, including for example tricuspid atresia with transposition of the great arteries. I also read that there is a classification system for the anatomic variations (listeners can see the script for additional details).

Preceptor: Correct. Can you walk me through how blood flows in this type of congenital heart disease postnatally? Start in the right atrium.

Student: Because of the lack of connection between the right atrium and right ventricle, there is an obligatory right-to-left shunt at the atrial level, either through a patient foramen ovale or an atrial septal defect, where blood flows from the right atrium to the left atrium. The systemic venous return therefore mixes with the pulmonary venous return before entering the left ventricle through the mitral valve. Usually there is also a ventricular septal defect, which permits communication between the pulmonary and systemic circulations. Mixed blood is pumped from the left ventricle to the aorta and from the left ventricle through the VSD and to the pulmonary artery **(Figure 1)**.

Preceptor: That's right. What might happen if the VSD was very small and the pulmonary artery was severely stenotic or atretic?

You hurriedly try to recall your first and second year fetal and newborn physiology lectures.

Student: Well, I think this is what you would call a duct-dependent pulmonary circulation. This would mean that there is inadequate flow through the pulmonary valve to the lungs. Blood therefore flows from the aorta through the PDA into the pulmonary arteries, which is why in this anatomy keeping the ductus arteriosus patent is crucial postnatally.

Preceptor: Wow! You have done your reading!

Student: So how did you find out that Sarah's fetus had tricuspid atresia?

Preceptor: Sarah was having her routine detailed anatomical scan completed, which is typically done between 18 and 22 weeks gestation, and a few things were noted, which

led to maternal fetal medicine and pediatric cardiology becoming involved. On the four-chamber view, the right ventricle appeared much smaller than the left ventricle.

Student: Do we know what caused this condition?

Preceptor: It's presumed that the tricuspid valve segments become fused during embryogenesis, forming either a muscular or fibrous atresia.[6] Why this occurs is unknown.[7] However, it has been found to be associated with heterotaxy syndromes and Schmid-Fraccaro syndrome,[6] though no specific genetic markers have been found to date.[8] We often describe congenital heart disease as being multifactorial in etiology.

Student: What kind of monitoring will Sarah require during her pregnancy?

Preceptor: Due to the difference in circulation with the placenta and lung physiology during pregnancy, compared to postnatal life, tricuspid atresia does not cause any acute circulatory problems prenatally and fetuses generally continue to grow and develop. We follow fetuses for growth of structures and assessment for neonatal risk stratification.

Student: Does special planning have to occur with regards to the birth?

Preceptor: For those women delivering an infant with known tricuspid atresia or other forms of single ventricle physiology, delivery at a tertiary care facility is recommended to avoid any transport-related morbidities and to allow the family and infant to remain close after birth.[9] A vaginal delivery is still recommended when possible. We advocate for delivery occurring at as mature a gestational age as possible, in order to allow for the best neurological development possible, given known associated neurodevelopmental complications. As you mentioned, we are also concerned about ductal patency. For this reason, following delivery, prostaglandin therapy is initiated to maintain ductal patency when needed, and an echocardiogram is performed to confirm the diagnosis.[7] Fortunately, in Sarah's case there is one thing that we know about her baby's physiology that makes immediate intervention less likely. Can you think of what that might be?

Student: The atrial septal defect?

Preceptor: That's right – because there is a patent atrial septal defect, we know that blood can move from the right atrium to the left atrium. Without this, her baby might require immediate interventional catheterization in order to create a communication between the two atria by balloon atrial septostomy.[7]

Student: Does Sarah's baby need surgery when it is born?

Preceptor: Sarah's baby will require a series of surgeries to palliate this congenital heart condition, but you'll learn more about that in your peds rotation.

Student: What do you mean by surgical palliation?

Preceptor: It is called surgical "palliation" because we are altering the primary anatomy, but we cannot fix it back to a normal heart, or in other words back to a normal two ventricle system. This does not mean palliative as in end-of-life care, nor does it mean that we are not aiming for best quality care and function.

Intrigued by everything you just learned about tricuspid atresia, you go with your preceptor to meet Sarah and participate in her MFM appointment.

* * *

2 months later you are in your pediatric core rotation and doing a subspecialty rotation in pediatric cardiology, which you are feeling much more confident about after your MFM rotation. You are rounding with the cardiology team, and your preceptor that week is the same pediatric cardiologist you met at the MFM clinic. Your team stops by the Cardiac ICU to see a baby boy who was born two hours ago with known tricuspid atresia. You go in with the team to assess the patient, and are delighted to see that the patient's parents are Sarah and her partner Coby. Sarah introduces you to her baby boy, whom they have named Jason. Your preceptor suggests you examine the baby and report the findings to her.

Student: On exam, Jason is cyanotic. He is sleeping comfortably in his bassinette. No clubbing is present, which is an expected finding, as clubbing is seen in older patients with long-standing cyanotic heart disease. On auscultation of his lungs, breath sounds are normal with no crackles or wheeze. His radial and femoral pulses are +2 and symmetric bilaterally. Inspection of his precordium is unremarkable. On auscultation, there is a single loud S2 heart sound with a grade 3/6 systolic ejection murmur most audible at the right upper sternal border. There are no extra heart sounds. Jason's liver is palpated at the costal margin.

Your preceptor examines Jason and nods to you.

Preceptor: I agree. Jason's exam is pretty characteristic for a newborn with tricuspid atresia. Inspection and palpation of their precordium is often unremarkable. The grade 3/6 systolic ejection murmur is suggestive of pulmonary stenosis. The duration of this murmur directly correlates with the amount of pulmonary blood flow, while the intensity inversely correlates with the severity of pulmonary stenosis and level of cyanosis (e.g. if there is no systolic ejection murmur, this suggests pulmonary atresia).[10]

Remembering back to a few weeks ago when we saw Sarah prenatally, what do you think is running in Jason's IV?

Student: Prostaglandin!

Preceptor: Correct. We are here to confirm the diagnosis by echocardiography, but I see that an ECG and chest x-ray have also been completed. What can you appreciate about Jason's chest x-ray?

Student: I can see decreased pulmonary vascular markings and a normally positioned heart of normal heart size.

Preceptor: I agree. What would you expect to see if Jason has increased pulmonary blood flow?

Student: Increased pulmonary vascular markings.

Preceptor: Yes. Would you expect to see a certain type of cardiac configuration on chest x-ray?

Student: Okay, I find this confusing. I was reading that all sorts of different cardiac configurations are supposed to be characteristic, like egg-shaped, boot-shaped, snowman-shaped.

Preceptor: That was a trick question, as this is a common misconception. While many configurations have been described, as you mentioned, there is no consistent pattern present in tricuspid atresia. In tricuspid atresia, chest x-rays are most useful in distinguishing between neonates with increased pulmonary vascular markings and those with decreased pulmonary vascular markings.[3]

Let's move on to his ECG. Can you interpret it for me please?

Student: Jason's ECG shows normal sinus rhythm at a rate of 160 beats per minute with a normal PR interval. There is a superior QRS axis, which is seen in tricuspid atresia. In other forms of cyanotic heart disease, the QRS frontal plane axis is inferior rather than superior, so the ECG can be used to distinguish between tricuspid atresia and other forms of cyanotic heart disease.[11]

Your preceptor and the pediatric cardiology fellow proceed with Jason's echocardiogram and the team goes to review the scan.

Preceptor: The newborn echocardiogram confirms a diagnosis of tricuspid atresia. In particular, we can appreciate the lack of a tricuspid valve opening, a hypoplastic right ventricle, normally related great vessels, a ventricular septal defect, severe pulmonary

stenosis and an atrial septal defect with flow from right to left. It is imperative for management and surgical planning that we note the following information on echocardiogram in general for any patient with tricuspid atresia: the ventricular septal defect size, the great vessel relationship and the presence and severity of outflow tract obstruction.[7]

Student: What about cardiac catheterization?

Preceptor: Cardiac catheterization is only performed prior to stage I surgical palliation in select cases when certain preoperative information is not available, as it can help to provide information about anatomy and hemodynamics that may influence management. It is routinely performed before stages II and III of surgical palliation.[10]

Patients may undergo cardiac catheterization for interventional or therapeutic purposes prior to initial surgical intervention. Can you think of a reason why?

Student: If they have a restrictive atrial septal defect. The goal of the catheterization would be to create a new atrial septal defect via balloon atrial septostomy to decompress the right atrium.

Preceptor: Correct. Patients who are duct-dependent may also have a stent placed in the catheterization laboratory to maintain ductus arteriosus patency versus a surgical intervention as a first step.[10,12] Interventional cardiac catheterization may also be the favoured approach in high-risk patients (e.g. prematurity, multiple congenital abnormalities, severe ventricular dysfunction).[10]

Student: Does Jason need any more intervention before surgery other than prostaglandins?

Preceptor: Medical management of a patient with a single ventricle is a fairly nuanced topic beyond the level of a 3rd or 4th year medical student. The goal in managing any patient with single ventricle physiology is to promote pulmonary blood flow and systemic blood flow in order to achieve relatively comparable levels of blood flow while maintaining ventricular function.[10]

We are meeting with the cardiac surgery team this afternoon to review Jason's case. Why don't you go and read up on the different surgical procedures for palliating tricuspid atresia?

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Through your reading you learn that surgical palliation of tricuspid atresia will generally take place over three surgical procedures ***(Figure 3****). The end goal of these three*

procedures, the Fontan circulation, is to separate the oxygenated blood from the deoxygenated blood and to have the heart pumping blood to the body.

Preceptor: Now that you've had some time to do some reading, how do we determine what type of primary procedure to do in a patient with tricuspid atresia?

Student: The type of primary procedure is determined by the amount of pulmonary blood flow, whether there is too much, too little, or whether it's just right.

Preceptor: That's right. There can be too much blood flow, too little blood flow, or blood flow that is "just right". So what happens in the setting of too little pulmonary blood flow?

Student: When there is too little pulmonary blood flow, such as in the setting of pulmonary atresia, the goal is to find a way to get more pulmonary blood flow, by keeping the duct open by stenting or replacing it with a synthetic duct.

Preceptor: Yes. A modified Blalock-Tasussig (BT) shunt is the most common primary procedure for neonates with coexisting severe pulmonary stenosis or pulmonary atresia **(Figure 3A)**. During this procedure a Gore-Tex tube is used to connect the subclavian artery and the ipsilateral (or same-sided) pulmonary artery.[10]

There is an excellent movie that you should watch about this by the way, called "Something the Lord Made". It's a fascinating movie describing Blalock, a white American cardiac surgeon and his African-American assistant, Vivien Thomas, who did many of the fundamental studies for the BT shunt himself, and actually stood behind Dr. Blalock, coaching him through his first surgery.

Student: Fascinating – I'm going to have to give that a watch!

Preceptor: Definitely. So what happens when there is too much pulmonary blood flow?

Student: When there is too much pulmonary blood flow, such as in the setting of a large VSD and no pulmonary stenosis, the cardiac surgeon can place a pulmonary band around the pulmonary artery, like a twist-tie, restricting the blood flow.

Preceptor: Correct, and when the amount of pulmonary blood flow is just right, such as in the setting of pulmonary stenosis with a small VSD, we call it the "goldilocks phenomenon" and no intervention is required until the Glenn connection at 6 months.

* * *

Jason is reviewed at the joint cardiology and cardiac surgery case conference that afternoon and is deemed a suitable candidate for surgery. He successfully undergoes the BT shunt procedure a few days later at 5 days old.

You review Jason with the pediatric cardiology team postoperatively during rounds.

Student: It sounds like Jason's procedure was successful. Why didn't they just do his operation right after birth?

Preceptor: It's important to wait for the drop in pulmonary vascular resistance, which occurs over a few days, otherwise the shunt would be unable to adequately supply blood to the lungs.

Student: Oh that makes sense. What will happen now as they wait for the next operation?

Preceptor: Following the first stage of surgical palliation, the interim stage is focused on supporting organ function and somatic growth. Management during this time focuses on pharmacologic therapy optimizing efficiency of the circulation, identifying concerning pathophysiology, and supporting growth.[7] Patients should also be monitored for cyanosis, and poor weight gain.[1] The patient will be discharged home and will be monitored closely as an outpatient.

Student: It was great to be able to see Jason through this first procedure!

* * *

Four months later you are on your surgical subspecialty rotation, which happens to be pediatric cardiovascular surgery. During your rotation you see Jason, now 6 months old, here with his parents for his Stage II palliation surgery (Bidirectional Glenn). You see the cardiology team visiting Jason on rounds and your old preceptor is surprised but delighted to see you.

Preceptor: This is great continuity of care for you to be seeing Jason receive the second stage of his surgery. What do you think about the timing of the surgery?

Student: From my understanding the second stage of surgical palliation usually occurs at 3 to 6 months of age, though performing Stage II prior to 4 months in patients at increased risk of interstage mortality is acceptable.

Preceptor: Correct. Which procedure will the surgeon do?

Student: A bidirectional Glenn.

Preceptor: What does the bidirectional Glenn achieve from a cardiovascular perspective?

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Student: The bidirectional Glenn starts the process of separating the circulation. This procedure will make it so that all the deoxygenated blood from the top half of the body now can enter straight into the lungs. This is achieved surgically by connecting the superior vena cava to the pulmonary artery. Completing this superior cavopulmonary connection prior to a Fontan (**Figure 3D-E**) increases survival and decreases perioperative and late mortality.[13,14]

Preceptor: Exactly. We will continue to follow Jason after his bidirectional Glenn to monitor for any transient hypertension, especially in the first couple of weeks postoperatively, as well as for any gradual decreases in oxygen saturation. Jason will be followed serially by the cardiology clinic, including assessment by echocardiography and ECG, to assess for any complications from the Glenn connection, such as Glenn obstruction. Should anything be suspected, then further investigations would be undertaken, such as cardiac catheterization, CT, or MRI.

Otherwise, life will look more normal for these children. They only need to come to the hospital every 6 months or so for check-ups and otherwise can lead a fairly normal life at home.

Jason has a successful bidirectional Glenn procedure and is discharged home with appropriate pediatric cardiology follow up.

* * *

Three years later you're a 2nd year pediatrics resident on your cardiology rotation. In clinic you see 3-year-old Jason, here for his pre-Fontan assessment. You are delighted to see Jason again, who has grown into an energetic, interactive little boy. You complete Jason's history and physical exam with a 3rd year medical student who has just started their pediatric cardiology rotation. While you are waiting to review with your staff, the clerk proceeds to ask you some questions about Jason's condition and his surgery.

3rd year clerk: I know that Jason is here to be assessed prior to completing a Fontan, his third and final surgery for surgical palliation of tricuspid valve atresia. When is this surgery typically done?

2nd year resident: Good question. So long as the patient has already received Stage II palliation, like Jason has, the timing of Stage III is not critical. Usually it is electively performed most commonly around ages 3-4 years.

3rd year clerk: What's the purpose of the Fontan surgery?

2nd year resident: The surgical goal of this procedure is to completely separate blue deoxygenated blood and red oxygenated blood. In this way, the blue deoxygenated

blood from the upper half of the body will flow through the Glenn at the top, and the blue deoxygenated blood from the lower half of the body will flow through the extracardiac Fontan. This is accomplished surgically by routing blood from the inferior vena cava to the pulmonary arteries.[7]

3rd year clerk: I understand that there's a lot of different ways that this can be done, is that right?

2nd year resident: There are many pros and cons to each type of procedure. In general, the extracardiac Fontan (**Figure 3E**) is the now the most common Fontan procedure. It involves placement of a prosthetic conduit between the pulmonary arteries and the inferior vena cava, and can be either fenestrated or nonfenestrated.[7] Fenestration is controversial, and the decision of whether or not to fenestrate is ultimately made by the surgeon. The purpose of fenestration is to allow for right-to-left shunting, improving ventricular preload and cardiac output at the expense of becoming cyanotic. Some studies have shown that this has resulted in shorter hospital stays and excellent survival.[15]

The pediatric cardiologist and pediatric cardiac surgeon examine Jason and review his most recent echocardiogram, cardiac catheterization, and ECG. They are pleased with his current clinical status and discuss him at multidisciplinary conference rounds, where he is booked for his Stage III palliation surgery, a non-fenestrated extracardiac conduit Fontan. Jason has a successful operation and has regular annual pediatric cardiology follow-up.

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There are many short-term and long-term complications and sequelae associated with the Fontan circulation. To learn more about these and see how Jason is doing in late adolescence, please refer to the third and final episode of this PedsCases podcast series. In our next episode we will be discussing hypoplastic left heart syndrome, another common cause of single ventricle physiology.

* * *

Recommended resources for learners

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Show Notes

Tricuspid Atresia Classification System

Tricuspid atresia is classified primarily by the position of the great arteries, and then into subgroups depending on the extent of blood flow to the vessel arising from the right ventricle. Type I describes normally related great arteries, Type II describes transposition of the great arteries, and Type III describes other complex anatomy. The most common type of tricuspid atresia, which is what is present in Sarah's case, is Type IB. This comprises about 50% of cases, and is characterized by tricuspid atresia with pulmonary stenosis/hypoplasia and the great vessels in normal anatomical position **(Figure 1)**.[6]

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Figures

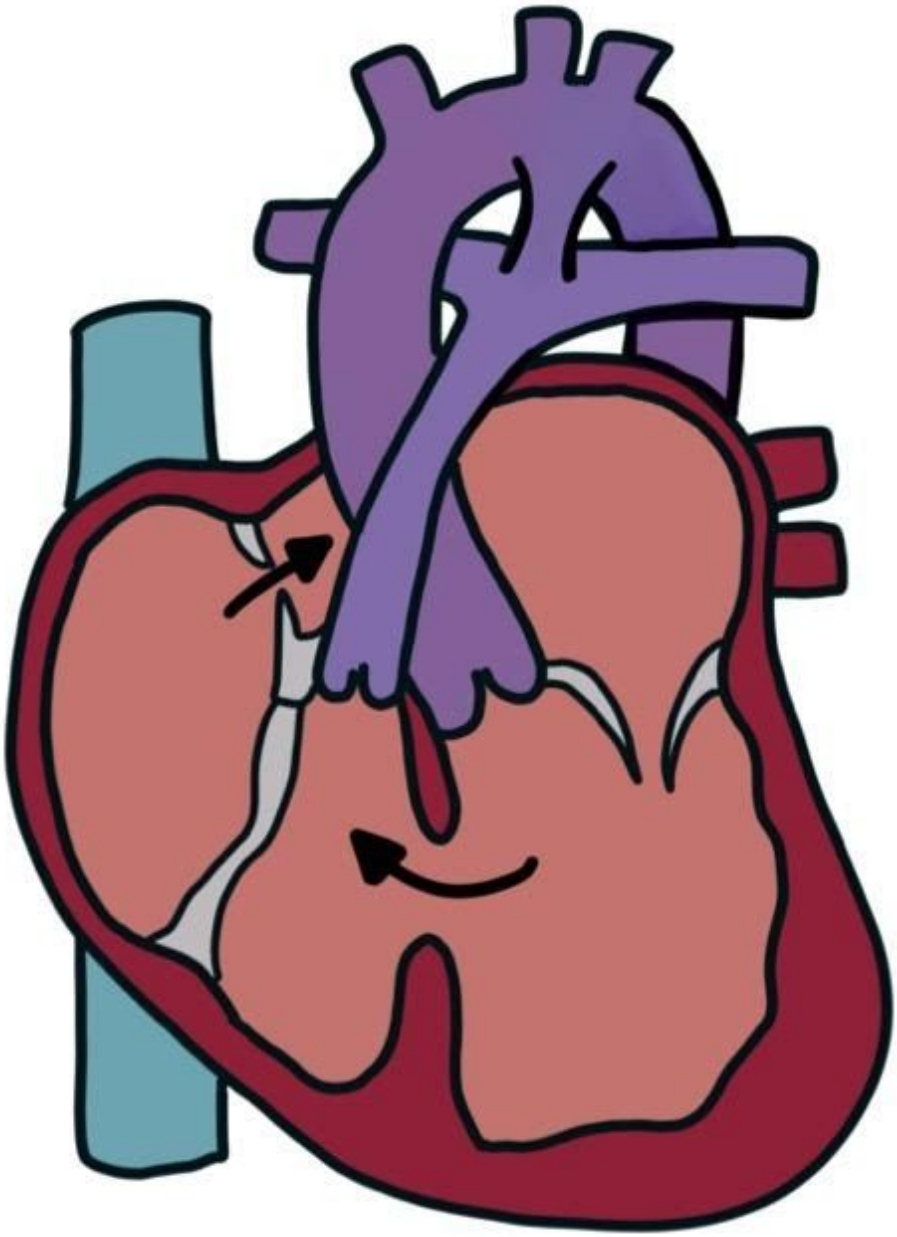

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Figure 1: Tricuspid atresia. Pictured here is the most common form of tricuspid atresia (type IB) with pulmonary stenosis/hypoplasia and normal anatomical position of the great vessels. Figure by S. Laguë.

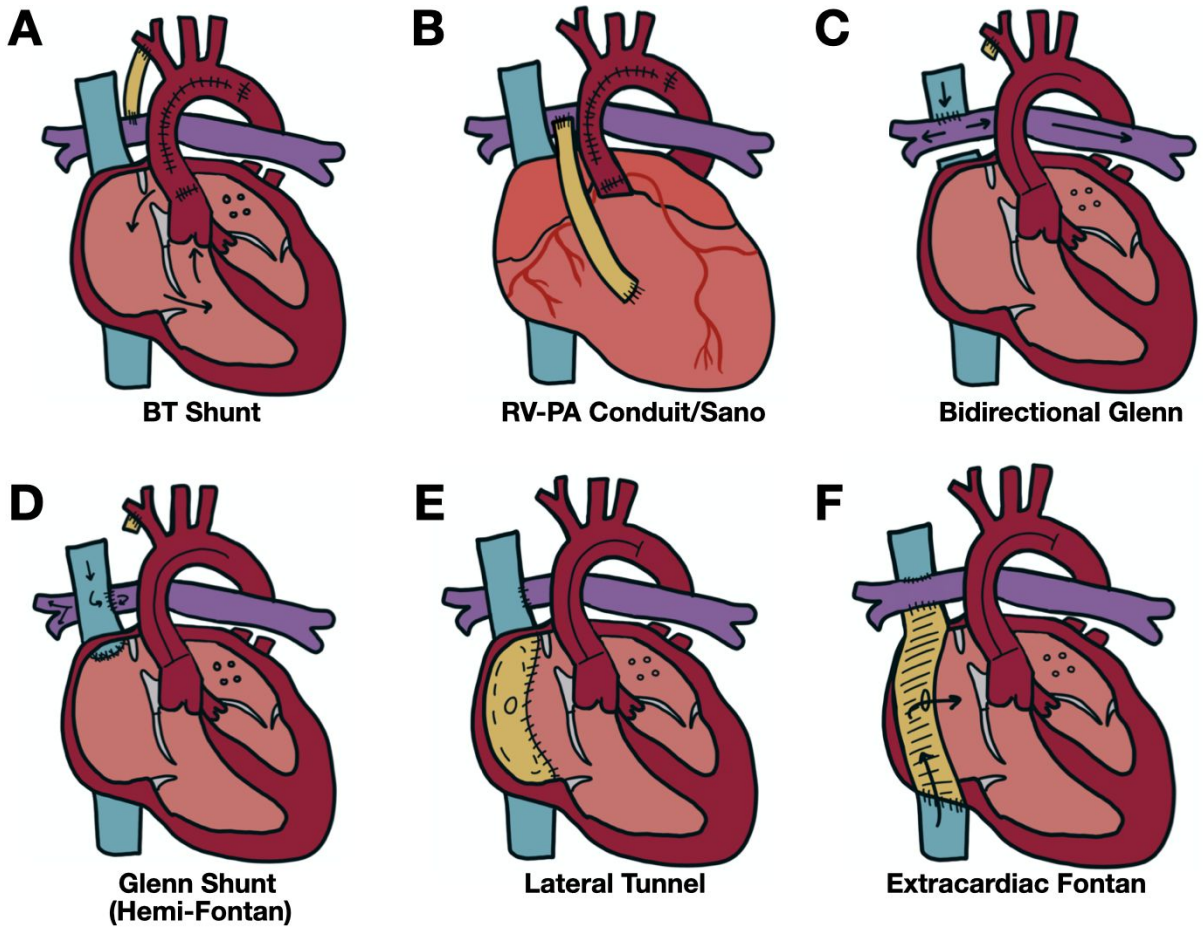

Figure 3: Surgical palliation repairs available for anatomically and functionally single ventricles, as shown in a heart with hypoplastic left heart syndrome (HLHS). These include the (A) BT (Blalock-Tasussig) Shunt, the most common primary shunt procedure; the (B) right-ventricle to pulmonary artery (RV-PA) conduit/Sano; the (c) Bidirectional Glenn), the most common second stage procedure, and its alternative, the (D) Glenn shunt or hemi-Fontan; and two forms of third stage procedures, the (E) lateral tunnel and (F) extracardiac Fontan (with or without fenestration). Figure by S. Laguë.

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