[MUSIC PLAYING]

CHRISTINA PISANI-CONWAY: Our objectives for today are to review first and second trimester obstetrical evaluation of the fetal heart to discuss institution specific performance of obstetrical screening ultrasound, to review indications for fetal echocardiogram, to identify genetic conditions associated with congenital heart disease, and review the most common abnormal cardiac lesions, and identify the components of an abnormal fetal echocardiogram, as well as discuss the post delivery management and surgical interventions for these lesions.

Congenital heart defects occur with an incidence of approximately 5 to 9 per 1,000 live births. According to a systematic review and meta-analysis of worldwide congenital heart disease, performed by van der Linde and colleagues, congenital heart defects are the most common malformations leading to early morbidity and mortality. Prenatal identification and management of fetal cardiac abnormalities is important, because congenital anomalies are the leading cause of infant death. And congenital heart disease accounts for 30% to 50% of these deaths.

Generally, the full spectrum of cardiac lesions seen in a postnatal population can be detected in the fetus, with the exception of some minor lesions such as the secundum atrial septal defects, or ASDs, which are less likely to be diagnosed in the prenatal period, and patent ductus arteriosus, or PDA, which is a normal fetal shunt.

Clinical indications for fetal echocardiography are often based on a variety of parental and fetal risk factors. However, most cases of congenital heart disease are not associated with known fetal and/or maternal risk factors, but rather, are often suspected at the time of an anatomic ultrasound survey. For women at low risk of having a fetus with congenital heart disease, cardiac screening is performed with a standard second trimester obstetrical ultrasound examination.

When risk is elevated above that of the general population, referral for fetal echo may be indicated depending on local resources, clinical settings, examiner availability, and results of a fetal cardiac screening evaluation. For women with average risk for an abnormality, a complete anatomy ultrasound is performed. For women with increased risk for an abnormality, an extended anatomy ultrasound is performed. There are many indications for an extended anatomy ultrasound. And there's significant overlap with indications for a fetal echocardiogram.

Indications for an extended anatomy ultrasound with regard to the scope of this talk include obesity, suspected maternal infections, pre-pregnancy diabetes, personal or first degree family history of congenital heart disease, multiple gestations, fetuses with known chromosomal genetic or other structural abnormalities, fetuses with disorders that can affect cardiac function, and fetuses with exposures associated with congenital heart disease, including medications and certain substances. For fetuses suspected of having an abnormal fetal heart at the time of a basic or detailed anatomic ultrasound exam, referral for fetal echo is indicated, as the risk of congenital heart disease is high in these patients.

This slide lists the cardiac views that are required by the American Institute of Ultrasound in Medicine and side-by-side with the cardiac views that are protocolized at Magee. And you can see some similarities. The four chamber heart view, the left ventricular outflow tract, right ventricular outflow tract, three vessel view, and three vessel tracheal view are collected as well as the cine clip, which is a brief video showing the active anatomy in live motion, typically for about 5 seconds. And that's an addition that McGee collects, which really contributes to better interpretation.

And this is a list of the views that are required in the video-- the views that are collected at Magee for the extended detailed anatomy. And as you can see, there is some mirroring in [INAUDIBLE] similarity. The extended anatomy ultrasound also adds in addition to what's collected for a basic exam situs, which is the right and left sidedness, aortic arch, the bicaval view with superior and inferior vena cava, and ductal arch, and interventricular septum.

Now, I'd like to take a few moments to walk you through the required images. I love this picture, because it demonstrates how close the planes that allow us to evaluate the fetal heart are in space. Their sixth view is required for a low risk complete anatomy ultrasound. The four chamber view is the most directly transaxial view of the fetal test, with the remaining views moving slightly more oblique.

The first view that we collect in complete basic ultrasound is the four chamber view. The four chamber view is on the lower left, and it provides us with a wealth of information. We can appreciate the size of the heart, its position in the chest, and its axis. And you can appreciate cardiac access by drawing a line from the fetal spine directly to the anterior aspect of the fetal chest dividing the chest into two halves. A normal axis is 45 degrees left of midline plus or minus 20 degrees. And we'll typically visualize the fetal heart in the left side of the chest and point it towards the left.

We can appreciate cardiac symmetry, contractility, and the presence of two distinct atrioventricular valves in this view. We can also appreciate that the septal leaflet at the tricuspid valve on the right side of the heart inserts slightly more apically than the mitral valve on the left side. And we can appreciate the pulmonary veins returning to the left atrium. We can also appreciate the apex forming nature of the left ventricle and the moderator band at the base of the right ventricle. And we can appreciate the descending aorta, anterior and slightly left to the fetal spine.

In the left ventricular outflow tract, we can appreciate the size of the aortic valve and mitral valve [INAUDIBLE] and the integrity of the interventricular septum. We can also appreciate that the left outflow tract arises from the left ventricle as well as other aspects that we've already reviewed in the four chamber view. In the right ventricular outflow tract on the lower right, we can appreciate the relative size, and position, and presence in some cases of congenital heart disease of the pulmonary artery, aorta, and superior vena cava.

Additionally, in the lower left, you can see the three vessel view with the main pulmonary artery bifurcation. Moving on to these additional three views that are collected as part of the complete anatomy ultrasound, the three vessel view with pulmonary artery bifurcation and the three vessel view with ductal arch allow us to appreciate the relative size and presence of these additional structures.

The three vessel tracheal view also allows us to appreciate the location of the brightly echogenic and cartilaginous trachea relative to the transverse aortic arch and the junction of the aortic isthmus and ductal arch. Adding color flow to these structures allows us to appreciate the presence, the directionality of flow, and the turbulence of flow, which may be abnormal in certain settings with congenital heart disease.

Additional views that we collect for the extended anatomy ultrasound include situs, which is the right and left sidedness of a fetus. This is a picture of one of our own babies and demonstrates how our ultrasound technicians communicate this information with the interpreting sonologists. This is a split screen of a picture across the fetal [INAUDIBLE], where we can appreciate based on the baby's position where the spine is, what side of the baby should be left and which side should be right. And this picture demonstrates that the fetal stomach and the four chamber are both located on the same side of the baby, which we expect to be the left side.

On this slide, we can see pictures of some of the additional views that are collected with an extended anatomy ultrasound. On the left, you can appreciate the aortic arch view that shows us that the aorta has no signs or no areas of narrowing. And we can see the origin of the brachiocephalic artery that divides into the right subclavian and right common carotid, the left common carotid and the left subclavian artery as they travel towards the fetal head.

The bicaval view on the lower right is a parasagittal view that demonstrates a similar caliber to the SVC and IVC as they enter the right atrium. And although this illustrated picture doesn't demonstrate it as well as it could, we can generally appreciate the right atrial appendage and the pyramidal shape superior aspect of the right atrium that helps us identify that chamber.

Additional views that we collect as part of the extended anatomy ultrasound include the ductal arch. The ductal can be distinguished from the aortic arch. And that has a more angular or hockey puck appearance versus the more candy cane shape of the aorta. And it arises more anteriorly in the fetal chest and provides no branches.

If we were to move just a bit from this plane, we could see the aortic isthmus join the ductal arch in the same Y shaped confluence that we see in the three vessel tracheal view. The interventricular septum is best assessed when the septum is perpendicular to the ultrasound beam, as you can appreciate in the right sided lower left short axis view of the fetal ventricles.

While the screening cardiac views are fresh in our mind, I wanted to take a moment to brag about Magee's rates of completion for screening views for the second half of 2019. I'm pretty proud of these numbers, and I'll orient you to this graph. On the left column, we have the anatomy view. And the reason that the aortic arch and the IVC and SVC are fewer in numbers is because they're only required for an extended anatomy ultrasound.

The next column to the right is the percentage of abnormal. The next column is the percentage of visualized and normal. And then added together is the percentage that these structures are visualized, and a call is made for each individual cardiac view. We make a call on between 87.3% and 91.8% of cardiac views on our first attempt at a complete or extended anatomy ultrasound. We need to re-evaluate between 6.5% and 12.7% of women to complete their OB cardiac screening.

With increasing maternal obesity and sonographers scanning at multiple satellite centers to facilitate more conveniently accessible high quality fetal ultrasound, this is the result of an enormous amount of effort on the part of our excellent sonographers, leading sonographer educators, administrative leadership, and quality improvement programs, and of course our team of physician sonologists led by Dr. David Kauffman.

Now, I want to take a moment and talk about with a ventricular echogenic intracardiac focus, or EIF for short. The EIF can be seen in the four chamber view, and you can see two pictures on the right hand side that demonstrate this, and also in the left ventricular outflow tract, and can be observed as early as the first trimester. Primarily, they're observed in the papillary muscles but not exclusively. And they can be single or multiple and located in either ventricle.

Academic foci are observed in 21% to 28% of fetuses with Down syndrome and 3% to 5% of euploid fetuses. So the majority of-- well not the majority, but a good proportion of babies with Down syndrome will have an observed echogenic focus and a smaller proportion of euploid babies will demonstrate this finding. But the majority of echogenic foci are going to be observed in euploid babies. And there's a higher prevalence in Asian mothers. So we see echogenic foci in up to 30% of Asian fetuses, most of which are going to be euploid.

Echogenic foci are thought to be related to microcalcifications and fibrosis in the papillary muscles or chordae. And they're believed to disappear later in pregnancy or postnatally. And I think the most important take-home point is that these echogenic foci are not associated with myocardial dysfunction or structural abnormalities. So there's no reason that our patients need to be concerned about these with regard to long term health for their babies if they're isolated, particularly if they've had aneuploidy screening.

So what do our professional organizations have to say about echogenic foci? This is an excerpt from the *Practice Bulletin* published in 2016 on screening for fetal aneuploidy. The likelihood ratio or the number of times that were likely of a baby to have Down syndrome with the presence of an EIF is between 1.4 and 1.8. And the presence or the prevalence of it in kids with Down syndrome and euploid babies is these numbers are very similar to the alternative source on the previous slide.

So I think this take-home point for management is that if this is an isolated finding, and the patient has not been offered aneuploidy screening, it's not a bad idea just to review options again, particularly if it hasn't been done previously. And if the patients already had an aneuploidy screen, and that result was reassuring with regard to Down syndrome, there's no further evaluation required. And you can present this finding as a normal variant.

I'd like to take a moment to talk about first trimester cardiac screening. We're always pushing the envelope with what we can see in the first trimester with the goal of providing earlier useful information without raising unnecessary alarm. If the fetal crown rump length measures between 41 in millimeters and 79 millimeters late in the first trimester, in the range where we would perform a nuchal translucency, you can imagine how small the fetal heart is at that point.

And having practiced in a lower resourced area north of Pittsburgh since my training and returning to Magee last April, I've definitely learned the value of early information and coordination. I typically did a pretty detailed first trimester ultrasound mirroring Magee's approach with an attempt to evaluate the four chamber heart view and a three view in the first trimester, for early identification and coordination of care, for patients at higher risk for congenital heart disease.

I'd like to briefly review a meta-analysis by Yu et al. published earlier this year in the *Journal of Ultrasound in Medicine.* This was a meta-analysis of 18 studies of over 26,000 fetal hearts examined between 11 weeks and 13 weeks and six days. They collected images both transvaginally and transabdominally with variable images collected across these studies, including the four chamber heart view, outflow tracts, three vessel view, and three vessel tracheal view.

The pooled sensitivity was 75%, and specificity was 99% for all congenital heart disease. And for major congenital heart disease, the sensitivity was even higher at 81.4%. And some kinds of congenital heart disease are progressive, including particular types the valvular stenosis and hypoplastic left and right heart syndrome. These appear more abnormal with advancing gestation and tend to be more obvious on the second trimester exam. And second trimester assessment is still necessary even if images can be collected in the first trimester.

So I wanted to take a moment in gratitude for Dr. Johnson and her pediatric cardiology colleagues including Jackie Wineberg, Dr. Jackie Wineberg, and Dr. [INAUDIBLE], Dr. Rachel Torok, and Dr. Nick McCaffery for their efforts in providing care to our patients, and Dr. Victor Morell and his pediatric cardiothoracic surgery colleagues as well as Dr. DaSilva, the coordinator of the Da Silva Center for Ebstein's anomaly.

So I wanted to take a moment to go through fetal echocardiogram indications. When do we need to send our patients to this incredible crew? We have two distinct places that we can look for fetal echo indications, and there's significant overlap between the two. There's a list of indications from the American Institute of Ultrasound in Medicine, which includes a suspected cardiac anomaly on OB scanning, suspected abnormality in cardiac function, fetal hydrops, persistent fetal tachycardia or bradycardia, as well as suspected fatal heart block, and frequent episodes or a persistently irregular cardiac rhythm.

Fetal indications also include a major fetal extracardiac anomaly, a nuchal translucency of 3.5 millimeters or greater, or at above the 99th percentile for gestational age, a chromosome abnormality by invasive genetic testing, or with cell-free fetal DNA screening, and monochorionic twinning.

Taking into account an additional indication listed by the American Heart Association, we can also consider fetal echocardiography if there's a systemic venous anomaly which would include a persistent right umbilical vein, left superior vena cava, or absent ductus venosus, and a greater than normal nuchal translucency between 3 and 3.4 millimeters. And the American Heart Association also adds an abnormality of the umbilical cord, placenta, or intra-abdominal venous anomaly.

Maternal factors that indicate a fetal echo include pre-gestational diabetes regardless of hemoglobin A1C, gestational diabetes diagnosed in the first or early second trimester, IVF including intracytoplasmic sperm injection or ICSI, PKU, phenylketonuria, either with unknown status or a periconceptional phenylalanine level greater than 10, an autoimmune disease with anti-Sjogren antibodies, SS-A, and a prior affected fetus.

Other maternal indications include first degree relatives of a fetus with congenital heart disease, including parents, siblings, or prior pregnancy, and first or second degree relatives with the disease of Mendelian inheritance associated with a history of childhood cardiac manifestations, as well as retinoid exposure and first trimester rubella infection.

Fetal echo can be considered with maternal factors including selected teratogen exposures-- for example, paroxetine, or Paxil, carbamazepine, or lithium-- and exposure to ACE inhibitors, as well as autoimmune disease with Sjogren antibodies without a prior affected fetus, and second degree relatives of a fetus with congenital heart disease.

I think the best take-home point from the American Heart Association list of indications for fetal echocardiogram include indications for which there's less than a 1% risk of congenital heart disease, and fetal echo is thought to not be indicated. This includes maternal gestational diabetes with a hemoglobin A1C less than 6%, and other maternal medications including SSRIs other than Paxil, vitamin K antagonists such as Coumadin-- although, an extended fetal survey is recommended-- and maternal infections other than rubella with seroconversion only, as well as isolated CHD, or congenital heart disease, in a relative that is not first or second degree to the fetus.

So I wanted to take a moment to review the Center for Advanced Fetal Diagnosis' general approach to coordination of congenital heart disease. Typically, genetic counseling with screening and/or diagnostic testing per patient preference is recommended. Fetal echocardiogram and pediatric consultation with follow-up is recommended, and growth ultrasounds every four weeks. Pediatric cardiothoracic surgery consultation is coordinated, as well as neonatology consultation typically later in the third trimester.

I wanted to take a moment just to review the genetic aspects of congenital heart disease. Many genetic abnormalities are associated with congenital heart disease, including aneuploidy syndromes, chromosomal dilation, and microdilation syndromes, single gene disorders with variable inheritance patterns.

I'm going to show you three slides with the genetic associations for congenital heart disease with the syndrome listed on the left and the cardiovascular anomalies on the right. These graphs are not meant to overwhelm, but they're meant to express the scope of genetic abnormalities that can be present in fetuses with congenital heart disease, many of which may have an independent relationship with prognosis and long term health.

We know our patients don't always have good recall of what type of genetic testing they've chosen and what is screened for with the tests that they've had. So with this knowledge, when your patient presents who's had low risk non-invasive prenatal testing, and they make a comment like, quote unquote, "there's nothing genetic wrong." You can feel empowered to review that there's many genetic changes associated with congenital heart disease that are not necessarily screened with non-invasive prenatal testing and know that collecting cord blood for microarray at delivery will likely be recommended.

And now, as we prepare to transition to a discussion regarding the most common congenital heart defects and their general management with Dr. Johnson, I wanted to take a moment of gratitude for what she offers to the women and the families that we care for. I pulled a few recent comments from the Children's Hospital website from her patients in July of 2020.

Dr. Johnson always goes above and beyond. She's amazing. And Dr. Johnson has been the absolute best doctor we've ever had. I felt so confident that my son was in the best possible place under her care. And I'm so grateful to UPMC for the experience. And we are grateful for what she does for our patients.

JENNIFER A. JOHNSON: Well, thank you very much. I really appreciate that. That's very kind. My job is to go through some examples. We'll go through the normal example and then some abnormals and then talk about the surgical and post op care. So objectives, we're going to review the specific abnormal cardiac lesions, identify the components of an abnormal fetal echocardiogram, and then just discuss post delivery and surgical interventions of each lesion. So I broke everything down into four chamber-- we'll get the three vessel view, and then we'll also look at outflow tracts as well.

So abnormal four chamber-- everybody's favorite, because these usually are nice and easy. And the identification of them is usually more on par. So here's an example of a normal for chamber view. So again, orientating ourselves, here is the spine. So this is the left, left atrium, left ventricle, right atrium, right ventricle. The ventricles are about the same size. And so are the atriums. So to us, that is normal, normal four chamber.

So we'll start looking at the abnormals. So differential diagnosis for an abnormal four chamber-- so what are the things that should be going through your head? So most common, atrioventricular septal defects, single ventricle lesions, hypoplastic left heart syndrome, tricuspid valve atresia, double outlet right ventricle with mitral valve atresia, double inlet left ventricle, and unbalanced atrioventricular septal defect.

Also, we get a lot of Ebstein's anomaly and tricuspid dysplasia due to our association with Dr. DaSilva and his advancement of the cone procedure. So we get a lot of patients from around the country and the world for this anomaly. So it's one that we will review since you will be seeing a lot more of them. And then congenitally corrected transposition of the great arteries, which can be a little tricky. So I thought that was a good one to review.

So we'll start with complete atrioventricular septal defect, the anatomy. So I think of an atrioventricular septal defect is that there's a hole in the top and the bottom, like the middle of the heart is completely missing. So the top hole is called a primum atrial septal defect. It can be different sizes. An inlet ventricular septal defect, again, can be different sizes. And I always like this picture, because it reminds us that the mitral and the tricuspid valve in a normal heart are isolated.

And in a complete atrioventricular septal defect, these valves have been put together and make one single valve. Anatomy wise, these can be tricky. Sometimes, the inlet ventricular septal defect is small. And I feel like a lot of times when we have our apex up in our four chambers, I definitely can make up an inlet VSD a lot of times on a normal heart. So we'll talk about what else to look for, not only in the four chamber, to diagnose an atrioventricular septal defect.

Genetics, as we've already talked about, there's a high association with Down syndrome. Any time we see an older mom with a complete AVSD, we always make sure genetic testing is at least provided or discussed. Prenatal monitoring, these patients we usually see depends on the family. You should see at least two times in fetal clinic just to make sure they understand the diagnosis, that genetic testing has been done or at least offered. And these patients, it depends if they have any other extracardiac anomalies. A lot of times, these patients can go to the newborn nursery or go to the NICU, because they have an extracardiac problem.

So here's the first four chamber example. So as you can see here, spine again, so left right. Here is your primum atrial septal defect. And here is your inlet defect. It's really nicely easy to see. You can see when the valve's open. They're not separated. They're one single valve, and the holes are nice and big. Here's a second example.

Again, just showing that there's a hole here that's the inlet ventricular septal defect and the primum. There's probably also a patent foramen ovale. That's why you get this. But I think the biggest thing is the primum atrial septal defect that really seals the deal. But sometimes-- and I have been fooled-- the inlet ventricular septal defect is small. So I know a lot of people talk about the AV valves being on the same level, which you can also do that in a normal heart.

So my go-to, because it's part of your protocol, is looking at the valves and making sure they're separated. So the other way you can look at and confirm is looking on the valves on [INAUDIBLE]. So you can see that as this clip goes through, the valve is one single valve. Do you see how they come together and instead of being two separate valves like we're used to, they're one big valve?

So a lot of times, if I'm doing a four chamber, I'm like, oh, is that an inlet VSD? I don't know if this is an AVSD. I will do this view. And if I see two separate AV valves, then I'm like, OK, win, these are normal. But if I see them together like this, then I know this is an AVSD.

This has really saved me a lot of times, because sometimes that inlet VSD is shallow. And this can always make you feel better. So I think if you-- especially, I think with a GE when the apex is up, I feel like you see a lot of fake drop out that looks like an inlet VSD that is not there. So by doing this image and showing that there's two separate valves or one single valve can help you determine if this is an AVSD or not.

So what's the surgical procedure? So these patients usually go home after the newborn nursery or the NICU, unless they have extracardiac issues. They usually have surgery on three to six months. I really like these patients to be as chubby as possible. My goal is around like 4 to 5 kilos before sending them to the OR, because the single AV valve is really the most important part of this repair.

So what our surgeons do is there's two types of repair. They can either do a two-patch or a single patch. So they're going to patch the ventricular septal defect and the atrial septal defect, either with one patch or two. And then the complicated part of the surgery is really cutting the single valve into two portions. And unfortunately, usually, the ventricles and the AV valves are not perfectly symmetrical. So a lot of times, it's at the surgeon's discretion how to cut these. Do they need to cheat over a little bit to the right to make the left a little bigger, since it's inherently small? But I really think the big part of this surgery is cutting these valves and getting that right.

So like I tell a lot of parents, it really all depends. And if for future surgeries if the valves go well and there's minimal leaking and minimal narrowing or stenosis, these kids might not need another surgery in their lifetime. If unfortunately, the left AV valve was small to start, these kids sometimes require AV valve replacements. It's usually in the left. And some of these kids, for some reason, will get subaortic membranes, which is a little membrane or outflow tract obstruction that needs to be respected later in life. It's hard to tell which kids will get those, but these kids can't have future surgeries in their lifetime.

Hypoplastic left heart, that's another single ventricle favorite and really nicely shown in a four chamber. So hypoplastic left heart syndrome, as the name says, the left ventricle is very small. Then the mitral valve is either atretic, or it has significant stenosis. And the aortic valve, which is not seen in this pathology specimen, is also very stenotic or atretic. Coarctation of the aorta is also associated with it. And then just the arch itself is hypoplastic and small.

When dealing with hypoplastic left heart, the biggest thing is I want to know if the patient has Turner's syndrome. Those patients sometimes can have worse outcomes. So we like to prep families. If their child does have Turner's syndrome, the outcome can be worse than just a baseline patient with hypoplastic left heart. So these patients, for me, it's very important to see them two to three times. Usually, we see them at diagnosis around 20 weeks. I like to see them at 28, because we can get really good pictures. And it gives us an opportunity to start looking at the pulmonary veins in the atrial septum.

Any single heart lesion and transposition, which we'll talk about, are all septal dependent. Especially here delivering at Magee where you're over two miles away from the Children's Hospital from my surgeons, from an hour, from ECMO, we really need to be on top of our game when dealing with hypoplastic left heart and transposition septums.

So these patients are seen at 28. And then again, the most important scan is 34 to 36. If you're only going to give me one scan, I would say just give me the 34 to 36, because you must look at the veins in the septum. As we've had about one or two a year especially last year, we've had patients with restrictive atrial septums, which you can tell by the pulmonary veins and which needed-- I organized a c-section at Children's.

The last patient we had was delivered last September. He came out, and his oxygen saturations were 3%. And I felt like that was anesthesia being very kind to me by giving him 3%. He was very blue. I sat there and imaged him in the OR, and there was no communication. So he's the patient that we are really lucky that we were able to determine that he had a intact atrial septum. And therefore, he got appropriate care.

So again, these patients, 34 to 36 weeks. If that's all you take away from this conference, that's the take-home point. These patients looking at the septum at 34 to 36 weeks. And again, these patients go right to the cardiac intensive care unit. That's where they need to be.

So here's an example. Remember, hypoplastic left heart is a spectrum. So this is an example of a patient who had mitral valve stenosis and aortic valve stenosis. You can see that the pulmonary veins are a little engorged, which would make me think, we really need to get these Dopplers and make sure there is not any issues that would be concerning for a restricted atrial septum.

So here's the LV. Since it was only stenosis of the left ventricle-- or stenosis of the mitral valve and aortic valve, there's actually an LV cavity. But as you can see, the RV does the major work for the patient. And then, here's an example of another hypoplast. So hypoplast can come in different forms. You can see that this is all just right ventricle. This is the left ventricle.

So this patient has mitral valve atresia and aortic valve atresia, because none of the left ventricle formed. Sometimes, you're thinking like, oh, it has to be a small left ventricle. Sometimes it's nothing, like this one. There's absolutely no left ventricle at all. This is still a hypoplast. So these are so the two examples you need to look for.

So hypoplastic left heart syndrome or any single ventricle, they're very tenuous. So these patients are delivered. Oxygen saturations are monitored closely. It's a continued balance of systemic and pulmonary blood flow. Just like everything in life, once your pulmonary resistance goes down, the lungs flood with blood. So everyone wants to take the path of least resistance.

So that means in these patients around two to three days of life, the pulmonary pressures drop. And all the blood flow wants to go to the lungs. The problem with that is that there's no blood that goes to the rest of the body. These kids become acidotic and can lead to cardiac arrest. So these kids go right to the cardiac ICU where they're monitored very closely.

The first procedure is done in the first week of life. It's called a Norwood procedure. It's got a high mortality rate, just for the procedure alone. So we're very thankful that we have a surgeon like Dr. Morell who does a really nice job on these procedures and has one of the lowest mortalities in the country. So in this procedure, the small little aortic valve is added to the big pulmonary valve. So now, this is what we call the neo-aorta.

So it's a small aortic valve with the coronary arteries, because you don't move them, added to the pulmonary valve, which now becomes the real aortic valve. Then, the coarctation that evolved in the arch is removed with patch material. So the arch is now made patent. And now that we've removed our pulmonary blood flow, a BT, or Blalock-Taussig, shunt is added. This is the crux of the issue. So this is a 3.5 millimeter shunt that gives the patient all their pulmonary blood flow. If this is kinked or has a clot in it, these patients will have an arrest.

So these kids are monitored in the hospital or the children's home down the street till about two to four months of age. Then, around two to three months of age, we start talking about taking down the BT shunt and moving towards a bidirectional Glenn, which is where or left superior vena cava is now added to the right pulmonary artery. This is a more stable blood flow. We always tell parents, this surgery goes really well. And you fly out of the hospital five days post op. And you're finally home. So patients are really excited when they finally get there Glenn.

After the Glenn, these patients are seen in cardiology clinic every three to four months, depending on any issues. And then around three to four years of age, we complete the single ventricle palliation by the Fontan procedure. That is taking now the blue blood from the legs through a conduit to the right pulmonary artery. The reason why we start with the head first in the first surgery or the second stage is because in babies' heads and upper extremities have the most blood flow. Then as we get older, the legs have the highest blood flow. So that's why it's done second.

The quote for hypoplast is about 60% survival to childhood. I think a lot of the data is hard, because some kids get heart transplants and other things. But I think the overall survival is a little bit lower than we all expected, but some of these kids do great. Some of them, unfortunately, do not. And it's really hard to pick as a fetus who's going to do poorly. So that's hypoplastic left heart.

And then we'll talk about Ebstein's anomalies. And these will all be coming our way. So Ebstein's anomaly in this pathology picture. This is the septal leaflet, and it's completely tacked on and displaced, and on the ventricular septum. And then, you have an inferior leaflet or posterior leaflet that's in the back that's usually very adherent to the muscle and doesn't really move.

Then, the one leaflet that everyone notices on the echo is the anterior leaflet, which is large and redundant, seen here-- or you can't really see the [INAUDIBLE]. But you'll see it in the echo. And it usually has a lot of just attachments to it, but it's [INAUDIBLE] like and has a lot of attachments. So this in itself, and then also, if you put color on, usually has some insufficiency.

It's associated with PFOs, ASDs, VSDs, RVOT obstruction. That's why we always, when you're dealing with an Ebstein's anomaly, we always want to know, is there pulmonary valve stenosis, atresia? Or sometimes it's functional atresia, because all the flow is going backwards. So the pulmonary valve doesn't have the opportunity to open. Patent ductus arteriosus, coarctation, left sided lesions, and corrected transposition. That's another one to look for when you see Ebstein.

Genetics, there's no high genetic abnormality in prenatal diagnosis. It depends on the kid. If there's significant cardiomegaly, and there's fetal distress, we see them maybe every four to six weeks. It just depends. These kids, depending on the cardiomegaly, will go to the cardiac intensive care unit.

So here's an example of a patient we had. So there's tricuspid dysplasia in Ebstein's anomaly. This one's more of a tricuspid dysplasia. But here, I think you can-- the end result and surgery is the same. So this is a recent patient of ours. You can see that the tricuspid valves, they don't touch. So that's a problem. So it's called tricuspid valve dysplasia, because there's not complete tethering of the anterior leaflet. So there's usually significant insufficiency that's causing this large right atrium, which is pretty big.

This is an example of Ebstein. So here's the left. Here's the right. This whole thing is your right atrium. Here's your left atrium. And then here is that septal leaflet that's completely tethered down. And then as you can see, here's your anterior leaflet. It's also significantly tethering. So you can see there's a huge coarctation gap. There's really not much flow going forward. It's all really going backwards. So you can put color on that.

And then this is always my favorite one. This is a clip from the Mayo Clinic. This is their worst Ebstein's they've ever had as a fetus. So here is your left atrium. This is your left ventricle. And this is your right atrium. Here is your Ebstinoid valve or Ebstein's valve. So these patients, when they get to be this size-- and we've had a couple-- sometimes they do not survive after delivery.

And it depends. I've had a couple that have significant cardiomegaly, and they did fine and made it to Children's not requiring ECMO till 24, 48 hours of life. So it really just depends. That's why patients with significant Ebstein's anomaly have their own post delivery protocol, which we practice with the NICU. So these patients also can be tenuous.

The cone procedure, which is performed by Dr. DaSilva, so it depends on what these kids look like. The patients I showed you in this clip, these all went for the single ventricle pathway, which is the Starnes procedure. Patients with not as significant congenital heart or Ebstein's can go for a cone procedure, which is what was developed by Dr. DaSilva around four to five years of age depending on how bad the valve is.

So congenitally corrected transposition, that's another one. This one, again, it's the ventricles are switched. Everything else is in the right spot, but the LV an RV are not. You can look for pulmonary stenosis VSD. These patients you see two to three times. Again, they can go to the cardiac floor or the ICU depending on what's going on.

So this is all about the moderator band, how you pick this one out. So here's your-- this should be your left side. So this should be the left atrium. This should be your ventricle. And you're like, wait a minute, where is the moderator band? There isn't one. So when you see a nice smooth wall, you really need to think about this. This is, again, another way you can look for your ventricles on [INAUDIBLE]. And you'll see that your left ventricle will be anterior instead of posterior. You'll see it in the papillary muscles.

So again, looking at the four chamber, you always want to look for that moderator band. Where is it? Here, you can also see that at a certain point that your great arteries are parallel. They won't be parallel like they would be for D-transposition, because the correct ventricles are going to-- or the correct aid-- or the correct semilunar valves are going to the ventricle that has the blue blood or red blood. So that's appropriate, but just the ventricles themselves.

So here, again, remember that this was the smooth wall LV. And then look at here. Your semilunar valve that's coming from the smooth wall left ventricle is a pulmonary valve. So that's always looking at the ventricle. What does it look like? Is there a moderator band?

And then also, this is a funny angle for a pulmonary valve to come off. So I think that's also another tip off, because obviously usually the RVOT comes off at an angle. And there's usually not continuity between the AV valve and the semilunar valve. So that's also a tip off. So these are tricky. I would say a lot of times, we get surprise corrected transpositions from the outside that aren't picked up. So these ones can be tricky. So today, when you're scanning, look for those moderator bands. Maybe you'll find one.

So corrected transposition, this has a multitude of surgical options. Double switch, which is the major procedure done before three years of life. You can just close the VSD. You can just monitor, or you can do a heart transplant. So these depend on the patient and what the family wants to do. So it all depends. But these are the four chamber options to look at.

And then abnormal three vessel view. So we like a good old sweep in cardiology. So this is a sweep. Here's the three vessel view. And you can see that the arch goes to the left of the trachea. So this is the left arch. So this has been really helpful. So things to look for-- transposition, tetralogy of Fallot truncus, and coarctation. We'll go over the first three.

Transpositions, this is different than corrected transposition. Now, our great vessels are the ones that are switched. The aorta comes from the right ventricle with the blue blood, and the pulmonary comes from the left ventricle with the red blood. So now, we have separate pulmonary and systemic circulations.

Things to talk to patients about-- abnormal coronary abnormalities. So that, we won't know until they're born. Ventricular septal defect and coarctation. We had one patient with a coarctation recently. The scary thing about this, just like the hypoplast, it's all about the evaluation at 34, 36 weeks. We have to look [INAUDIBLE] atrial septum. It can be tricky.

I know if you come to fetal clinic, I talk about jump ropes and looking for flow and trying-- I always tell parents, I can tell you if you need a septostomy after delivery. I can't tell you if you won't need one. So a lot of times, we look to make sure it's not completely intact, because if it was completely intact, we would deliver this patient at Children's.

If there is definitely a hole there, we would just do the transposition delivery protocol, which is to get the cath lab and the patient over to the cath lab within two hours after delivery. So these kids, again, for transposition of hypoplast, all I care about is 34, 36 weeks. If you come at 32, we make you come back. So it's really important that you come at that time.

So here, again, is a nice four chamber. And as you watch it move, here's the left side. You notice, wait a minute, it's branching. So here's your left ventricle. And you see that the semilunar valve branches. That is definitely a tip off that this is transposition. And then you can see that the semilunar valve coming across from the RV does not. I think with this sweep is the most important to diagnose for a transposition.

And then, the three vessel view, which is all about location, so location. So here's the moving clip, and here's the still. So you can't see the SVC. But remember, again, here's the left, because there's pointing. So here's right to left. So the order is correct. The SVC should be here. It's aorta, pulmonary, and this is your descending aorta. But the aortic valve comes first.

Remember, the aortic valve should be behind the pulmonary. So that's always your tip off in the three vessel view. I personally don't diagnose transposition in the three vessel view. It's all in that four chamber sweep. But you can tell from the first image, if you do that sweep, you can tell if a patient has transposition or not. So I think that's a very key sweep to do.

Again, surgical repair, done in the first week of life. It's an arterial switch. So our surgeon cuts the semilunar valves above the level of the valve. As you switch the coronaries, which is the coronary transfer, is the trickiest part. And then this doesn't show up, but they drape the branch pulmonary arteries above the aorta in the LeCompete position. And that's the arterial switch. These kids usually also do well. They can have branch pulmonary stenosis. As long as the coronary transfer is done well, they do really great. You would never know they had congenital heart disease later in life.

Tetralogy of Fallot, another conotruncal abnormality, again a large ventricular septal defect with an overriding aorta and a small RVOT tract. These patients have a high risk of DiGeorge. And we usually see them two to three times, depending on how small the pulmonary valve is. This patient can go to the NICU if the pulmonary valve's normal size, the cardiac ICU if it's atretic or small.

So here is a three vessel view. Again, you can see that the pulmonary is way smaller than the ascending aorta. So this is a nice tip off. These are helpful over the three vessel view. I like to use, again, looking for sweeps. So you can see quickly that there is a hole here and the aortic valve overrides it on the sweep.

So then when you do your slower evaluation, you can see that the aortic valve overrides the ventricular septal defect. And if you saw from the previous image, the pulmonary valve came from the RVOT but was good size. So we would consider this a pink tet. So this would be a good one. The one shown in the three vessel view is probably more of a blue tet. It will probably need surgery earlier or not in the neonatal period.

And then the [INAUDIBLE] this is always my reminder when you do the three vessel view, when you think of tets, you have to think of right arches. So again, here you see, and you see that the arch goes to the right of the trachea. So remember, when you are imaging a tet, do the three vessel view and look for arch sidedness. I think about 20% of the time you will get a right arch. It always brings me joy. There's no real difference. It's just that supposedly those patients have a higher risk of DiGeorge seen in a couple of [INAUDIBLE] studies.

So these patients usually have surgery. They do well after delivery. If it's a pink tet, they have surgery three to four months of age, which consists of either a valve sparing repair where they can save the valve and just patch above and below, or they do a transannular patch, which unfortunately ruins the valve. So you get free PI. Those patients need future pulmonary valve replacements in their lifetime. But these kids overall do well, except for you always have to be careful of DiGeorge that can have multiple extracardiac issues.

Truncus arteriosus, again, another conotruncal defect. This time, we just have one big semilunar valve and a big VSD. These patients, again, these have a higher risk of DiGeorge. I think 35%. We usually see them two to three times. They'll go to the cardiac ICU as well. I always tell fellows when they're scanning and they-- tip off-- and they see one vessel, you either need to find the a pulmonary artery or you have a truncus on your hands. So when you see something like this, you have to think of truncus.

Once you see this, you have to prove to me where the branch pulmonary artery is. Is it pulmonary atresia? Is there a pulmonary somewhere with branches? Or do the branches come off the ascending aorta? So here, you can see that the branches come off the ascending aorta. So here's your truncal valve. I think this is the right, and this is the left. So it's a type 2 truncus. So once you figure out that it's a truncus, you need to tell me where the branch PAs are to prove it. And here's another example. You can see them nicely coming off the truncal valve separately. So it'd be a type 2 truncus.

These patients have surgery in the first month of life usually in the first week of life. We usually don't let them sit, because they have a high risk of having pulmonary hypertension. So what our surgeon does is they close the ventricular septal defect. So the truncal valve becomes the aortic valve. And then they put an RV to PA conduit that connects the branch PAs that they take off the ascending aorta. Obviously, this conduit would need to be replaced, usually replaced after two years with the first one. And then after that, it just depends, every two to five years for the rest of their life.

And then the last section is the abnormal outflow tract. So here's a normal LVOT, and this is a normal RVOT. You can see nicely the pulmonary valve and the RPA and ductus. So we'll start differential diagnosis or looking at aortic and pulmonary stenosis. So we'll start with pulmonary stenosis. We've had a rash of those recently.

So the pulmonary valve, as you can see, doesn't open as well. It should open like a field goal. But here the leaflets are tethered, causing increase in aliasing and flow and velocity. And we usually see these two to three times. Again, the last appointment at 34 weeks is the most important. It tells us, can this go to the NICU, or this go to the cardiac ICU?

So this is a recent patient of ours. You can see this pulmonary valve is stuck. It doesn't move very well. And I think the color just brings it home. So you've got pulmonary valve insufficiency. And then you can see that it aliases. As this goes through, the main pulmonary artery is dilated. And then you look at the velocity, and it's 3. So for a normal semilunar valve, it should be less than 2. It should be more closer to 1 and 1/2, 1 if it's gestational age. So this is pretty significant. Usually, whatever the last velocity is is what it will be as a neonate. So this patient did have a balloon after delivery.

Intervention is cath as a balloon valvuloplasty, which is usually done in the cath lab. Some kids require surgery, but very rarely. And these kids usually do well. Depending on the balloon and the valve, sometimes they need multiple cath interventions in the first year of life. Some kids need to go back to the surgery later in life if the valve is really poor.

Aortic stenosis, the same thing. These patients, we watched probably a little closer. It just depends on what the LV looks like, if it's having function problems and things like that. So here's an example of a patient you can see as the flow aliases through. And this patient had a velocity close to 2. So this patient had mild aortic valve stenosis.

And then this is a patient of ours that had significant aortic valve stenosis. You can see there's barely any flow going through here and that the left ventricle is very thickened, echo bright. And this patient went down the single ventricle pathway. And this is another one of our patients who had significant aortic valve stenosis. You can see the valves barely move. And this left ventricle was dilated with EFE. And this patient went down the single ventricle pathway as well, like a hypoplast.

So these patients, usually we try to do a balloon valvuloplasty if we can, so the mild, moderate. Those patients with really bad LVs usually go for a single ventricle pathway. And so usually the pathway is balloon valvuloplasty, surgical valvotomy, worst case scenario a Ross. And then those with really bad LVs go down the single ventricle pathway. So I think we've got one minute to spare. So I'll answer questions and go from there. Thank you.