[MUSIC PLAYING]

LIZBETH LANFORD: Good morning. I'd like to thank the organizers for allowing to speak on one of my favorite topics-- echocardiography. So when I'm asked to give a talk, my typical first step, as I think most of us do, is a literature search. And so I did a literature search.

And I came across this paper by Dr. Morell, "Nikaidoh Procedure: How I Teach It." And in this paper, he gives us a nice roadmap. I think this was specifically for Nikaidoh procedure, but I think it's useful for patients with transposition VSD, PS because it's similar underlying anatomy.

So to quote, "Before the start of the operation, it is important to review the preoperative studies to have a clear understanding of the cardiac anatomy, including spatial relationship of the great vessels, size and location of the VSD." As an imager, I would add a little S there because we want to make sure that there aren't multiple VSDs.

"Coronary anatomy, size of the pulmonary annulus." and that's certainly important. If the pulmonary valve is severely hypoplastic, it's unlikely it's going to be amenable to any kind of surgical intervention. However, as an imager, we also look at some pulmonary stenosis, the leaflets, the morphology, "Presence of abnormal AV valve attachments, right ventricular size."

I added one more, because this is what I do in my thought process when I review these images and when I prepare images to review with the surgeons. I try to visualize how, on my images, I can get the LV to the semilunar valve. The cases that I'm going to show, we actually baffled the LV to the aorta. But sometimes in these children, we have found if the sub-PS is clonal septal tissue, it can be potentially resected. And maybe you're going to want to baffle the LV to the pulmonary valve and do a switch.

So I'm going to review the anatomy of two complex transposition patients. So the first one, I'm going to start with the subcostal sweeps to give a general overview of the anatomy. These were both referral patients, so I don't have any newborn images.

This is a 10-month-old with d-TGA, small VSD, pulmonary stenosis, status post a balloon atrial septostomy. So this first sweep here on the left is a subcostal four-chamber or coronal. It's going to be head, feet, right, left. So we're going to be sweeping from posterior to anterior.

Here's the RA and here's the LA. And there's the ASC from the balloon atrial septostomy. And as we sweep, we'll see the LV, and then over here is the RV. This is the aortic valve. I'm going to let that play again. And you see the pulmonary valves here, and the aortic valve is here.

So another thing that I like to do is, from the subcostal views, get the relationships of the great vessels, because your parasternal and other views are in the plane of the heart and not in the anatomical plane of the body. And the way I do that is, I put my cursor over the pulmonary valve. And as we come up anteriorly, you notice that the aortic valve actually is minimally leftward of this pulmonary valve.

So the aortic valve was anterior and leftward in this case. It's still the underlying anatomy of d-TGA. But again, that would be a nomenclature argument. And the other thing you notice in the sweep is, I haven't really seen a big VSD go by.

So moving on to the subcostal sagittal view-- so it's going to be head, feet, anterior, posterior, and we're sweeping right to left in this case. And so the first semilunar valve you'll see is going to be the most rightward. I'm just going to let it play. So here's your pulmonary valve, and here's your aortic valve.

So they're not pure side by side, but certainly, they don't give you the feeling of an anterior-posterior relationship. There's good-sized ventricles. And on the end of that sweep, you see what looks like a VSD and this small gap up here. So again, we're starting to be suspicious that the VSD in this case is not that large.

So moving on to the next view, this is an off-axis parasternal apical. And the reason I chose this view, even though it's a little bit off axis, is this is as big as the VSD as we ever could image. Here, it's 8 millimeters, which probably for a nine-month-old is probably OK. But that's not what you're going to want when you're 15 or 20.

And here, if you look, the thin line here is the pulmonary valve leaflets. And below that-- and I don't know why. It's a little bit fuzzy, that is actually conal septum that is deviated posteriorly, and causing the majority of the obstruction at subpulmonic stenosis.

For the fellows out there, there's an advantage to being a fellow. This is a great picture. And if we'd stuck in the standard views, we never would have got it.

All right, moving on to the parasternal short axis view to evaluate the VSD some more and the subPS and PS. And this is a beautiful short axis view, right atrium, right ventricle. So here is your aortic valve because it's anterior, it's a little bit leftward.

And here's your pulmonary valve. And the things that jump out at me is that this is a hypertrophied RV with good function. But most of these infant children have pretty significant RV hypertrophy since we don't tend to do these types of repairs in the neonatal period.

Here is the VSD, which again does not look very large. It is below both of the semilunar valves. And if we look over here on the still frame, it's a beautiful example of continuity between the aortic and the pulmonary valve.

So the anatomical definition, whether you use outlet doubly committed supracristal And you can see that the membranous septum is intact. And then there's this bar of tissue, which is the conal septum, which is causing some of the pulmonary outflow tract obstruction.

So it was easier for me to show on this particular patient. This is the pre-op TEE, how you would envision the LV-to-aortic pathway. So for people who aren't familiar with TEE, this is the LA. Sorry, that's not the LA. Here's the LA.

The mitral valve-- here is the pulmonary valve, which is in continuity with the mitral valve, transposition, dilated MPA, and the aortic valve. And here is the septum. And as you follow it up, there is the VSD.

So it's definitely an outlet VSD. But when you start to draw your path this way, certainly, the aortic valve is close, but it's not very wide. And I envision, I would not want my systemic output to go across that hole. Per Doctor Morell's request, we got the pulmonary valve annulus 1 centimeter. The z-score was minus 1.1, so actually within normal limits.

We always look for coronaries. And this child, this is the parasternal short axis view. So again, another look at that aortic valve, which is slightly anterior and leftward-- the pulmonary valve, which I think is actually bicuspid. You see a large coronary coming off on the right side.

And actually, even by 2D, you can see the bifurcation. And if we look at the color, the blue jet is the right coronary. And then this is the left coronary coursing anteriorly over the aorta. There should never be a great coronary coursing over the anterior great vessel.

So you know, you have a coronary anomaly. And if you look there's a branch that goes superiorly on this one. But that's rightward, which will be your LAD. And there's a blue branch here, which will be your circ. So we were very suspicious that this was a single right coronary, which is not advantageous for surgeries that involve moving the aortic valve or re-implanting coronaries.

So the intraoperative findings was a single coronary that originated from the right-sided sinus and crossed anterior to the aortic root. There was pulmonary stenosis that appeared to be at the valve. And it says supravalvular level. I left that in, but I still wonder whether that was a typo and was meant to be subvalvular. The VSD was somewhat small, doubly-committed outlet-type VSD.

So this patient had a Rastelli procedure with a 14 millimeter PTFE valve conduit between the right ventricle and the pulmonary arteries, a VSD enlargement, and atrial septal defect closure. And although I'm asked to discuss the assessment, I couldn't help it. I have to put in a few post-op pictures to show you what they look like, because there's no post-operative imaging shown on the conference.

So this is that subcostal sagittal view again. So it's head, feet, anterior, posterior. This is the SVC and RA. So we're going to sweep from right to left.

And as you sweep, here is your LV, and here is your RV. And you have to do it backwards because the aortic valve was up here. The outflow tract between the aorta and the LV is widely patent.

And because that was a little bit hard to see on that sweep because of the orientation. I did take some still frames for people. So here is a still frame that shows you-- sorry, I'm trying to get the other one to stop moving.

Here is the aortic valve. Here is the VSD patch. Here is the crest of the septum. So you can certainly see that VSD is much larger.

Also, you can see that the septum would have lined up this way vertically. So all of this volume was in the RV, and is now on the LV side. Now, most of these patients have good sized RVs because they've been having a systemic RV pressure for months.

But in this case, I don't think it was a problem. But I do think that this RV cavity, when we look at the RV, is smaller. It's still probably normal because the RV was so big to begin with.

And then looking at the parasternal long axis view, you can see how the inflow from LV now has to go anteriorly, almost a right angle, and then superiorly. So the normal LV outflow tract would have gone horizontally. Now this blood flow has to go anteriorly to get to that anterior great vessels. And again, just looking at that.

And you can also see how the VSD patch is attached on the right side. And I'm assuming that's to make the LV outflow tract bigger. But this child actually did quite well, and did not have a long hospital stay for his transposition issues.

Moving on to a slightly different patient, this was a 19-month-old, so already a bit older than that first patient, with double outlet right ventricle-- I can debate that one, TGA, subpulmonic VSD and subpulmonary stenosis, status post a PDA stent, balloon atrial septostomy, and a 3.5 central shunt. So again, starting in that subcostal view, where we're sweeping from posterior to anteriorly, here's your LV. You start to see pulmonary valve here, some stuff.

And then we come all the way up to the RV and the aortic valve. So it's a good-sized RV, good-sized aortic valve, but again, trying to look mitral valve. There's extra tissue here.

There's the pulmonary valve, and then the aortic valve. And then the only other thing to think about is, in this case, when I put my cursor over the pulmonary valve, the aortic valve is pretty much straight anterior. So this is more of an anterior-posterior relationship than the other patient.

Sweeping from right to left, pulmonary valve, aortic valve. And again, you see the pulmonary valve and the aortic valve. But we really can't see much about the subpulmonic area.

So we're going to move on to the apical four-chamber view. You can see, this is quite a dilated RV, a lot of hypertrophy, the disadvantage of waiting till 19 months. But when you look at the mitral valve, there's some attachment or tissue near the crest. And as you watch, there's some dropout here. So maybe there's a VSD there.

Again, this was a fairly complex patient. This child had never had any intracardiac surgery, but already had MR, at least mild. And just to remind people, if you're going to make the left ventricle the systemic ventricle, you need to make sure that it's prepared. This MR gradient was over 70, so we thought the LV was prepared.

Let me skip to this next picture to actually show you better the LV outflow tract. By the way, this is the argument. This is an anteriorly for the mitral valve. Here's the pulmonary valve, and they're in continuity.

So you can argue whether this was d-TGA versus whether this is double outlet. But if you look closely, here are your pulmonary valve leaflets. Here is all this extra tissue, which I've been calling stuff, or accessory tissue that is in the subpulmonic area. But it doesn't actually appear to be connected to that anterior leaflet.

And here, you start to see the VSD. And here is your aortic valve. And I don't think it's a great pathway. You're going to have to go through that tissue, and then just to look quickly.

What was important about this is that there are [INAUDIBLE] lots of attachments to the crest of the septum. And it's hard to tell how much of it is that accessory tissue, whether there's a mitral valve chordae attaching there, because I always worry about mitral valve function after resecting this tissue.

And as an imager, it's very hard for me to tell what's primary versus secondary chordae, what can be cut. But there's also a cord that comes here and attaches the crest of the septum that is from the tricuspid valve. So we've got the concern of mitral valve and tricuspid valve chordae in that VSD.

This is looking at my path to the VSD to the aortic valve. And again, you can see there's a lot of stuff in the middle. Plus I don't see a large 2D defect. The flow is getting through, but it's very worrisome for how you would baffle that.

And then this is just another look at how the baffle is going to look. Here's your LV, here's your RV, here's your aorta. So in addition to all the stuff in the middle, the aortic valve is fairly far away. And your baffle would have to run all the way across here, walling off a portion of the RV, but also giving you a large portion of the wall that doesn't contract VSD patch.

This is just to show, this child had normal coronaries normal right, normal left, speeding up. We're lucky enough to have one of the pediatric 3D probes. And so this is just a nice picture from the pre-op. I'm going to focus on this one. This is 90 degrees.

Here's your pulmonary valve. Here's your LA. Here's your anterior leaflet. And since that tissue is mobile, but not as mobile as the valves, you can see, this is that subpulmonary accessory tissue that goes from the coronal septum all the way down to the crest of the septum. So it looks like it's going to be resectable without injury to the mitral valve.

Let's see if I can just do it this way. OK, operative findings-- left ventricular outflow tract obstruction that was partially muscular in the subpulmonary area, membranous with a membrane that extended from the coronal septum down to the crest of the ventricular septum, that was somewhat parallel to the anterior leaflet of the mitral valve, which is, I think, exactly what that 3D image looked like. VSD was quite distal to the aorta.

The imager in me can't resist from telling you, it's not important for the surgical thing. But there was a small cleft in the anterior leaflet of the mitral valve. There were some abnormalities of the mitral valve.

So this patient underwent a Nikaidoh procedure, aortic translocation with a 14 millimeter RV to PA conduit. I actually was lucky enough to do both these pre and post-op TEEs. And this is utterly amazing to me still.

So here is your LA. Here's your anterior leaflet of the mitral valve. Here is your LV. Here's the native septum. Here's your VSD patch.

And here's your translocated aortic valve. There is no tissue there. It is wide open. I'm still amazed that worked.

Here is a 3D picture of it, which I think is nice, because again, it shows you with depth that there is no tissue there remaining. And the aortic valve here is off the screen. Again, not part thing, but the mitral valve worked just as well, if not a little better, post. So there was really no damage to the mitral valve with the surgery.

[SIDE CONVERSATION]

And then this is the last post-operative picture. So this is a transgastric view. It looks a lot like a subcostal, so it's going to be head, feet, right, left. So here is your RV with the RV-to-PA conduit.

Here is your septum. Here is your VSD patch. Here is your LV and aorta.

And you can see that the LV-to-aortic tract is wide open. It has more of a typical orientation. It doesn't have a bend to it. And you can also see that this RV cavity has been well preserved, compared to that other patient that I showed. And I did put some color in LV to RA, and RV to PA were widely patent.

And then my key takeaway from all of these images, when I was reviewing to prepare the study, is that each of these children is unique. They have subtle abnormalities that will determine which is the optimal surgery. And you have to carefully assess all of the features.

I am transthoracic echo person and TEE person. I do think that this is the area of multi-modality. I think this is the area of 3D.

When it gets better, it will be very helpful. But I will also give the caveat, because I have seen it, especially with one of my own patients, that there can still be subtle abnormalities in the OR that lead to a change in the surgical plan. But we do our best to try to get as much information to plan the surgical approach. Thank you.

[APPLAUSE]