REPAIR OF A COMPLEX CONGENITAL CARDIAC DEFECT
CHILDREN'S HOSPITAL BOSTON
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00:00:14
ANNOUNCER: Over the next hour, live from Children's Hospital Boston's cardiac OR, see cardiac surgeons repair a complex congenital cardiac defect. Each year the pediatric cardiac surgery program provides surgical care to approximately 1,100 patients, including more than 700 cases of open heart surgery that require a cardiopulmonary bypass and 400 closed cases that do not require a bypass, with extraordinary success rates. In just moments, the cardiac team will present this complex congenital cardiac case and answer your email questions. OR-Live makes it easy for you to learn more. Just click on the "request information" button on your webcast screen and open the door to informed medical care. Now let's join the doctors.
00:01:02
EMILE A. BACHA, MD: Hello. I would like to welcome you to this live webcast. My name is Emile Bacha. I'm one of the cardiac surgeons at Children's Hospital Boston and associate professor of surgery at Harvard Medical School. The theme today is LV rehabilitation in borderline left ventricles, something we have extensive experience with here at Children's Hospital Boston. I would present the patient that's currently in the operating room where Dr. del Nido has already started the surgery, and then we'll go to the OR and look at the setup and look at the echo images. Can we go to the slides, please? So AW is currently eight months old. He was prenatally diagnosed at 22 weeks gestational age with severe aortic valve stenosis. Of note, his mother is an obstetrician. He underwent a technically successful in utero aortic valvuloplasty and post-procedure had severe aortic regurgitation, which completely resolved, however, in late gestation. He also had moderate to severe mitral valve regurgitation and a restrictive atrial septum, which led to high left atrial pressures. He was born at 37 weeks gestational age. He underwent a neonatal aortic valve dilation, which resulted in a very good relief of great input, moderate to severe aortic regurgitation. At about two weeks of age, he then underwent a surgical aortic and mitral valvuloplasty, including extensive EFE resection. And he -- at that point, the noncoronary cusp of the aortic valve was augmented with a pericardial patch. Post-procedure there was only mild AR and mild AS and mild MS as well. He was discharged from the hospital at six weeks of age -- can we go to the next slide, please? -- and has done well at home. He's currently taking all his feeds orally. On physical examination, he is in no significant distress. His weight is 6.5 kg, but he's not growing well. The echo -- the surface echo showed a mixed aortic valve lesion with moderate aortic stenosis and moderate aortic regurgitation with mild to moderate MR and moderate mitral stenosis. He has a dilated left ventricle with mild dysfunction. At catheterization, which was done a few days ago, his LV pressure was normal. His mixed venous saturation was 70%. He had a mean gradient across the mitral valve of 5-6 mmHg. He had a 50 mmHg gradient across the LVOT with mild to moderate MR and moderate AR. Can we go to the next -- to the video, please? So this is an aortic root injection showing the amount of AR, which is a moderate grade. Now we will go to the operating room, where Dr. del Nido has already opened the chest and dissected out the cardiac structures. Can we?
00:04:01

PEDRO J. del NIDO, MD: Thank you. We're now doing an epicardial echocardiogram to try to confirm the transesophageal diagnosis of mitral stenosis and also to determine the exact mechanism of the stenosis. It looks from the transesophageal and the 3-D echo that most of the problem is really with the posterior leaflet, which is probably the most common cause. I know it's a very thickened and frozen posterior leaflet, which you can see on the 3-D echo image is a major source. The anterior leaflet does move and it does coapt reasonably well. There's mild mitral regurgitation, but most of the gradient is just from this posterior leaflet, and that's part of what we're going to address today. The other problem is the aortic valve -- just switching over on the epicardial imaging to look at this aortic valve. And we're beginning to see it there at the apex of the pyramid on the echo. And the arrow is actually pointing the leaflets out. That's a pretty good image there of the left and right coronary leaflets, and then you can see the noncoronary leaflet, which is on the left of your image, basically is immobile. This was what was initially repaired with a small pericardial patch when he was a neonate, and what's happened I think is the pericardial patch is probably thickened and immobile now and is a source of central regurgitation. You can probably apply some color there at some point and show the -- there's the regurgitant jet. Dr. Marx is going to show you some of the preacquired images that will show you in more detail some of the anatomic features that we are interested in.
00:06:05
GERALD R. MARX, MD: Thank you, Pedro. So Juan-Carlos Muniz, our fourth year fellow, and I acquired some transesophageal echocardiographic imaging. And we also did some epicardial three-dimensional echo imaging with Dr. del Nido, and we're just going to go through some of those images right now. We're going to start off first with our transesophageal echocardiographic images. And first is the short-axis image. And basically, what it will show is -- slow it down a little bit. You can see right here, this is the short-axis view of the aortic valve, so here, juxtaposed to the interatrial septum, is the noncoronary cusp. Then from this image from below, this will be the right and the left coronary cusp. And you can see there was good mobility and motion of these cusps, but where surgery had been done on the posterior noncoronary cusp, you can see it's quite immobile and very, very poor excursion. So from this image, we then went to placing a colorful jet on it. And you'll see that when we do the color compare, again, if you look at the image to the left you'll see right in here, you'll see sort of a gap. And when you look at it on the color flow image that's superimposed, you can see that the aortic regurgitation jet certainly emanates from the region of this deficient noncoronary cusp posteriorly. Now, we go on to look now…at the aortic valve -- sorry, mitral valve. And you can see that the leaflet tips are quite a bit thickened. Here's the anterior leaflet and thickening of the distal tips. But most notably, if you look at the posterior leaflet, again, there's thickening of the distal tips, but you can see there's poor excursion with a lot of thickening posteriorly, which oftentimes is related to endocardial fibroelastosis and thickening, which Dr. del Nido will go in and sort of excavate and really increase the mobility of that posterior leaflet. When we put the color flow image over it with the superimposed imaging, you can see that there's just mild mitral regurgitation, and you can certainly see that there's a significant stenosis with turbulence distal to the thickened mitral valve leaflets. Now, at this point in time we're going to turn our attention to some epicardial three-dimensional echocardiographic imaging. This is a young child, and so we used a small pediatric probe. And the advantages of the small pediatric probe is you really get improved near-field imaging and you get really -- well, we think it's very good resolution. So if I can just show you the -- sorry, I rotated it here. This is going to be the region of the left ventricle right here. Let me just rotate it down a little bit. And what's nice is that this is a live image in which there was no gating done, but what the surgeon can do is he's scanning the heart in three dimensions since we can see it on the field at the same time. And again, if I use my pointer, this is the anterior leaflet. And these are going to be those accessory chordae that Dr. del Nido had talked about that was preventing the excursion of this anterior leaflet. If I just slow it down and I go through this

frame by frame, what I want to point out is in this three-dimensional image now, is you can see the severe thickening of the anterior leaflet. This is that thickened excretion as you look from the apex to the left ventricle up towards the mitral valve. And we'll go to another image here for a second. And this is now a long axis parasternal. This is what would be similar to a long-axis parasternal view. Again, this is live imaging using the live imaging and not the full-volume, where you get a larger field of view. But you can be imaging and looking at these images as you're scanning. And again, because you save it you can look at it and post-process it, but if you'll allow me to just, again, just rotate this around.
00:11:02
EMILE A. BACHA, MD: Jerry, how far is Pedro in the case at this point?
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GERALD R. MARX, MD: What's that?
00:11:06
EMILE A. BACHA, MD: How far along is Pedro in the case?
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PEDRO J. del NIDO, MD: We're going to be cross-clamping in about a minute.
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EMILE A. BACHA, MD: Okay, because -- okay.
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GERALD R. MARX, MD: So here you can see the stenotic mitral valve from above in the left atrial view. And for time's sake, we'll quickly move on. We'll go on to some other images here.
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EMILE A. BACHA, MD: You already cross-clamped the smaller one?
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GERALD R. MARX, MD: And this is the full-volume view that we got of the aortic valve. And most notably, you can see that this is the retracted posterior leaflet. This is the right coronary cusp, and this is the left. And here's where the AR would emanate from right here. And then when we look at the three-dimensional color flow status set with it, you again can see that the AR emanates specifically from that region.
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EMILE A. BACHA, MD: And that's the cusp that was augmented previously with the pericardial patch.
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GERALD R. MARX, MD: That's exactly right. And if we just go to one last image, this is a long-axis parasternal view but now oriented as if looking out the left ventricular outflow tract. Here's the anterior mitral valve leaflet. Here's the circumferential view of the left ventricular outflow tract. And you can clearly see a significant subaortic membrane, concentric subaortic membrane, below the aortic valve, which Dr. del Nido will also excise at the time of the surgery.
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EMILE A. BACHA, MD: So I would say that the main benefit of 3-D in this particular case versus 2-D was to see that there is extensive tissue in the subaortic area that we weren't quite sure how much we were going to find, but certainly the 3-D is showing a large amount of tissue in the subaortic area. Can we go to the surgical images, please? Because there are issues with the patient having previous aortic regurgitation with respect to delivering of cardioplegia, and you cannot fill the root adequately with cardioplegia. And what I assume Dr. del Nido is doing, he is currently making the aortotomy. And it looks like the heart is immobile and has stopped. I'm not quite certain whether he's going to want to give cardioplegia directly into the coronary ostium --
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PEDRO J. del NIDO, MD: Actually, we're pretty good.
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EMILE A. BACHA, MD: You're okay? All right, good.
00:13:29
PEDRO J. del NIDO, MD: So we're beginning to cut down towards the aortic valve. We're beginning to see it now. The thing that's striking is that the noncoronary leaflet is really quite thickened and mobile, and that's what we need to repair. So we're just now starting to get exposure to this area.
00:13:55
EMILE A. BACHA, MD: So he's putting stay sutures on the upper side of the aortic incision to expose the aortic valve. Typically we like to put the stay sutures at the tips of the commissures. You can see that this is a redo case, having had his first surgery at two weeks of age. Event is in place via the right superior pulmonary vein that you can see in the right side of the picture.
00:14:42
PEDRO J. del NIDO, MD: So we want to get a pretty good view of not only the leaflets of the aortic valve, which you can see here. This is the right coronary leaflet where my suction is, and my forceps are in showing you the left coronary leaflet. As you can see, both of these are thickened, but probably the most striking thing is the fact that the noncoronary leaflet is really adherent and thickened almost not well developed at all, which is what we had seen on the original procedure. The pericardial patch, which as a newborn was very tiny, looks like it's all adherent posteriorly. So right now I'm just going to mobilize the two leaflets a little bit better by doing a commissurotomy in the intercoronary commissure, which will help mobilize the two leaflets that are actually functioning quite well. Fine forceps. This leaflet is quite thick, and I'm going to try to make an incision in it to try to thin it out.
00:16:07
EMILE A. BACHA, MD: So you're thinning the leaflets right now.
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PEDRO J. del NIDO, MD: Yeah, just the edge of it because it's really quite thick. And part of the problem why it doesn't coapt is just that it's just a little too thick.
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EMILE A. BACHA, MD: Right. And this is the left coronary cusp that you're working on.
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PEDRO J. del NIDO, MD: Left coronary cusp, exactly. This is not normal leaflet tissue, obviously.
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EMILE A. BACHA, MD: So it looks already more mobile right there.
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PEDRO J. del NIDO, MD: Yeah, we started at the plane of tissue that you can see is just --
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EMILE A. BACHA, MD: You're on the aortic side of the cusp.
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PEDRO J. del NIDO, MD: Yes. Okay, that's good. That'll help with this left leaflet. We can do a little bit more, but right now I'm going to look at the right coronary leaflet.
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EMILE A. BACHA, MD: Pedro, I'm going to address one of the questions here posited by the audience: "Can you repair rather than replace the valve?"
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PEDRO J. del NIDO, MD: Absolutely.
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EMILE A. BACHA, MD: Especially since this is the second time.
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PEDRO J. del NIDO, MD: Yeah, absolutely. The plan is to do the repair. The major part right now is to thin out these leaflets so that they can coapt well. They already coapt reasonably well, but if these came down, especially this leaflet came down by thinning this portion out,

then I don't need to augment the noncoronary leaflets so much. The other thing you begin to see is if you look below the valve, you're beginning to see the subaortic membrane right in this area, and we'll address that in a minute.
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EMILE A. BACHA, MD: So just to develop the answer to that question a little bit more, we would not be taking this child to the OR with a plan to replace this valve. The plan here is to repair the valve. And we've had fairly good results with mid to long-term with valve repairs in this setting. And the principles of the aortic valve repair are being well shown here by Dr. del Nido, one of them being the thinning of the cusps as he's doing so that you increase the mobility of the cusps in addition to the commissurotomy that he did initially. That was the first maneuver that he performed. Now again, he's developing that plane, trying to separate the real cuspal tissue from the membrane and tissue overgrowth that's over it.
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PEDRO J. del NIDO, MD: It's almost like ectopic tissue that's growing on the leaflet, and once you get the plane going, then you can just remove it all pretty well. This clearly is not like a normal leaflet that would be essentially transparent. These leaflets are typically quite thick at this age, and thinning them out really makes quite a difference as far as their excursion and how well they coapt with each other.
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EMILE A. BACHA, MD: Here you're at the commissure and you're thinning out the commissure.
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PEDRO J. del NIDO, MD: On the inner side, on the aortic side, again, to allow mobility of this leaflet.
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EMILE A. BACHA, MD: Right. This is the left-right commissure.
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PEDRO J. del NIDO, MD: Exactly. So I haven't started to work on the really defective leaflet, which is the noncoronary, yet. The idea is to get these as thinned out as possible.
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EMILE A. BACHA, MD: The added advantage of doing all this is that it'll give you a much better view of the subaortic area once you've thinned out and mobilized those leaflets.
00:21:03
PEDRO J. del NIDO, MD: Okay, that's pretty good. That's much better. So we're now getting leaflets that almost fall on each other, we're getting redundant, so we're starting to get to where we want to go, which is normally if you look at a valve, the leaflets fall on top of each other and automatically just close the gap. Here this leaflet has been fused, so we're going to create -- re-create the area of the sinus of Valsalva.
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EMILE A. BACHA, MD: So one question that came is what is the risk of regurgitation during commissurotomy.
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PEDRO J. del NIDO, MD: During or as a consequence of commissurotomy?
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EMILE A. BACHA, MD: I think so.
00:21:46
PEDRO J. del NIDO, MD: Yeah. As long as you still have well-supported leaflets afterwards -- in other words, the incision is only really in the commissure itself and not into the leaflets -- then it should work fine, it should work very well. In fact, it usually does. So here we're beginning to develop the noncoronary sinus of Valsalva. As you can see, I'm opening in the area.
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EMILE A. BACHA, MD: Is that pericardium that you're starting to see?

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PEDRO J. del NIDO, MD: Yes. Down here at the tip of my knife is the mitral valve, anterior leaflet of the mitral valve. And I can see already the abnormal secondary chord to that, which we're going to address in a minute.
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EMILE A. BACHA, MD: So this is all tissue overgrowth that has grown over that pericardial patch that was placed seven months ago.
00:22:43
PEDRO J. del NIDO, MD: Exactly. But once you sort of begin to really mobilize it, it's actually not too bad. It's -- my goal is to try to get rid of all the overgrowth and try to get back to as much of this normal -- this child had an aortic valve leaflet there that was torn, so much of that leaflet should still be here. And it's amazing how if you're able to leave some leaflet tissue, it actually begins to function a little better. Can you grab that? That's good. So as you can see, this child had essentially an obliterated sinus of Valsalva as a consequence of this. Now, if we let this go you can already see that now this area which used to be completely plastered is now beginning to come together almost like a leaflet. Now clearly, this part is fused here. This commissure's fused right there. And this leaflet is still quite thick, so we're going to thin this out and open this commissure.
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EMILE A. BACHA, MD: And for the surgical audience that may be wondering about myocardial protection, we give -- it's a one shot 20 cc per kilo at 4 degrees centigrade of high potassium magnesium solution. So we give one dose of cardioplegia for the entire duration of the myocardial arrest. So now you're opening the left noncommissure.
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PEDRO J. del NIDO, MD: Exactly. And what I'm going to try to do is get towards the -- all the way down towards the anterior leaflet of the mitral valve. We'll address the rest of that when we get to the subaortic membrane, but clearly that's part of the problem here. So can you grab the right leaflet for me?
00:25:03
EMILE A. BACHA, MD: So it's almost a little bit like a subaortic membrane that's grown onto the valve with tissue that's also underneath the valve.
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PEDRO J. del NIDO, MD: Yes. Exactly. So we already have a pretty good orifice of this aortic valve, as you can see. Hopefully you can see this.
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EMILE A. BACHA, MD: Yeah.
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PEDRO J. del NIDO, MD: We're beginning to see now a little ring directly below the valve. That's a subaortic membrane. And if you go a little deeper, I'm just grabbing the secondary chords on the anterior leaflet of the mitral valve. Those secondary chords are causing part of the problem here. See, these are the secondary chords, and they're fused. I'm going to just hook one of them. See, these are fused to the base of the -- sorry, to the middle of the leaflet. And I'm just pulling it through. And those are the things that we're going to divide. You can see that directly below are the real edge chords. You probably can't see that very well, but we'll see it much better once we resect the subaortic membrane. So we'll work on it through the LV outflow track, and obviously we'll also work on it through the left atrium.
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EMILE A. BACHA, MD: Pedro, the next question I will answer. It's somebody's daughter had open heart surgery in 1995, transposition of the great arteries. Doesn't say whether they had a switch operation or not, but she has a bicuspid aortic valve that remains. "Can a bicuspid valve from birth remain working throughout your life without needing repair?" The answer is yes, it can. Some people live to be 80 with a bicuspid aortic valve, but it's very variable. A fair amount of people with bicuspid aortic valve end up needing surgery in the

fifth to sixth decade of life because usually these valves calcify more rapidly and readily than normal valves. Having said that, it doesn't mean that your daughter will need surgery just because she has a bicuspid aortic valve.
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PEDRO J. del NIDO, MD: So we're just continuing the thinning process of these leaflets.
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EMILE A. BACHA, MD: I think here the instruments are key in that one difficulty in doing this, as Dr. del Nido is working, is to grab, is grabbing the tissue. And we really haven't found the right forceps yet to do that, especially once you -- the bulk of the tissue is excised and you're down to excising the rest of it, it gets harder to grab. And he looks like he has a good hold on it here, and he's using micro-scissors to excise this. Of course, there's a risk of cusp perforation, and so you have to know at what level you're at. Another question is, "How long is it appropriate for the heart to be under anesthesia?" I assume -- well, a person can remain under anesthesia for many, many hours -- days, in fact -- but in terms of the heart, I assume that the person is asking how long can the heart be stopped? And the answer is we've become much better at myocardial protection with improved cardioplegia, and we have cardioplegic arrest time, so when the heart is stopped during which we have to do the work that we need to do inside of the heart of several hours sometimes. Of course, we try to minimize the time as much as possible, but it's more important to do a good job as Dr. del Nido is doing here rather than rush through the procedure, which is what they had to do in the '60s and '70s when they did not have good methods of myocardial protection. So now we have the luxury of being able to concentrate on what we do and try to do the best job possible without having to worry that the myocardial function would be impaired post-op. Another question is causes for the growth of the subaortic membrane. The short answer to that is nobody really knows. It may be a flow phenomenon, flow turbulences, but nobody really knows what causes these membranes to go.
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PEDRO J. del NIDO, MD: So Emile, we're more or less complete with the thinning process. It's quite a bit better. This leaflet may have to be augmented with a little piece of pericardium right here. This is the noncoronary leaflet. I was trying to avoid that, but we may have to augment that a tiny bit. And then we're going to switch over to the mitral valve. So I'm just going to do a little more work on this noncoronary leaflet now and see if we can get it just to where we want it.
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EMILE A. BACHA, MD: I mean, for sure, compared to when you started, the effective orifice of the valve has increased quite a bit and the mobility of the valve cusps has increased by quite a bit. One can see that just when you move the cusps.
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PEDRO J. del NIDO, MD: Yeah, it's looking much more like a normal leaflet and how a normal leaflet would function. So now if I let it go, even if I pull the commissures apart, the leaflets come together. And that's what we're looking for because the natural process of what we call coaptation is occurring. This leaflet is still quite thick and we'll probably still thin it out, but I think we're going to work on the mitral valve now.
00:31:45
EMILE A. BACHA, MD: Okay. I'm going to answer another question: "Is this surgery being done to prevent having to perform the Fontan surgery?" A few years back, this patient may have received a Fontan procedure, yes. So the short answer would be yes, we're trying to move the border of when a patient needs to have a Fontan procedure, and that is why we try very hard to rehabilitate those borderline left ventricles, so those ventricles that are borderline incised, we're trying to push them back into the two-ventricle territory versus the single ventricle Fontan territory. So Dr. del Nido is now opening the right atrium and is going to --
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PEDRO J. del NIDO, MD: We're just lateral to the left atrium now. And so we're going trans left atrial in this procedure because this baby has in fact quite an enlarged left atrium. Not all of them do. In fact, the majority do not. But in this situation, what it does is allows it to avoid an incision on the right side. So now we're just going to begin to expose the mitral valve.
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EMILE A. BACHA, MD: So again, those [unintelligible] are placed for exposure. You can see the color of the endothelium in the atrium, which is white, and it's probably also some endocardial fibroelastosis tissue that is not supposed to be there. And it's the same process. You can see the whitish color of the inside of the left atrium there. Again, that's an abnormal color.
00:33:51
PEDRO J. del NIDO, MD: It should be normal -- or almost a single cell layer thick. And in babies with this kind of disease, it is quite thick and quite abnormal, and we think -- one more -- we think it's part of the problem. Take one more. All right.
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EMILE A. BACHA, MD: Somebody's asking what the congenital defect is. We're dealing here with critical aortic stenosis that had a fetal aortic balloon valvuloplasty followed by a neonatal balloon aortic valvuloplasty followed by mitral and aortic surgical valvuloplasties with good result with achievement of two ventricle repair. The patient had also EFE resection at that time, at the time of the first surgery. And the left ventricle has grown well. We're now dealing with some residual defects of mixed aortic valve disease, AS and MS, as well as mild to moderate MR with mild MS.
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PEDRO J. del NIDO, MD: And one more of those. So we're beginning to get the mitral valve into view here. And what we're going to focus on is the posterior leaflet primarily. We're going to address some of the anterior leaflet thickening through the aortic root, but now we're going to look at the posterior leaflet. So this is anterior leaflet here. This is the leaflet that's relatively mobile. This is the posterior leaflet, and as you can see, it is quite adherent to the free wall. There are -- most of these children have multiple papillary muscles. There's the primary one, which is anteroseptal here, posterior medial here. But this child also has a number of additional papillaries, and in fact, some that don't even attach to the leaflet itself. Small malleable retractor. So it is that rigidity, or immobility, that is causing a good bit of the stenosis. So if I lift this posterior leaflet, I can see that it is really quite thick. The chordal attachments are both at the edge, which here's a chord that's on the edge, but also there are chords behind. And these chords behind are a good bit of the problem. So this leaflet edge we can thin out and get mobility out of it, but we also need to deal with the problem that's primary here, which is the secondary chords that are pinning the posterior leaflet onto the wall, preventing it from coapting. So to address that, what we're going to do is separate the leaflet from the wall for over an extensive area -- 11, please? -- and then reattach it.
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EMILE A. BACHA, MD: Just to remind the audience that the initial surgery on the mitral was EFE resection and resection of secondary chordae at the time when the patient was a neonate. So we're going parallel to the annulus, to the posterior annulus here.
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PEDRO J. del NIDO, MD: Coronary scissors.
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EMILE A. BACHA, MD: Dr. del Nido is doing that because access to those secondary chordae that he has to cut is much more difficult if you went through the orifice of the valve. So by opening up that area --
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PEDRO J. del NIDO, MD: You can actually see the secondary chordae very well. This is the free edge that we've just cut, and these chords shouldn't be here. They shouldn't be attaching to that edge of that leaflet. So we're going to remove them to allow that leaflet to become much more mobile. Fine stapler? So I'm just going to now lift this leaflet up so that we can get exposure to the underside more easily. And what we want to do is all these matted secondary chords, we need to remove them because they're what is preventing this valve from really moving. So I'm just going to start dividing their attachments.
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EMILE A. BACHA, MD: And here you're starting at the ventricular aspect of the mirror leaflet, right? The ventricle edge.
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PEDRO J. del NIDO, MD: Yes, yes. It's --
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EMILE A. BACHA, MD: I mean, the septum or -- not the septum, but the posterior edge.
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PEDRO J. del NIDO, MD: Exactly.
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EMILE A. BACHA, MD: And it's fair to say that the chordae that are going to the free edge should be preserved and all those who go to the body below the free edge are actually able to be resected.
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PEDRO J. del NIDO, MD: Exactly. What it does is not only does it allow the leaflet to move, but it also changes the length of that leaflet. Because when these chords get matted together, they in fact shorten -- not just shorten it from the hinge point from the papillary in that direction, but they also shorten it from side to side. The other forceps -- the other fine ones. This tissue is really quite abnormal, very similar to what we see in the aortic valve. It's almost cartilaginous. It's very difficult to grab and cut. But as you can see it, you can remove it pretty well.
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EMILE A. BACHA, MD: And again, it's about developing a plane between what's normal tissue and what's abnormal tissue. And once you have that plane developed, you can follow it.
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PEDRO J. del NIDO, MD: Yep. And so you can see that in this section, at least, I've removed some of the secondary chords. The primary chords are still attached to the edge of the leaflet, but it's gotten quite a bit longer already, and this is what we're going to try to accomplish throughout the length of this leaflet. Usually once you get this plane started, it's quite a bit easier to continue it.
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EMILE A. BACHA, MD: And just what temperature are you at, Pedro, 22 degrees?
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PEDRO J. del NIDO, MD: Body temperature? Twenty-eight.
00:42:55
EMILE A. BACHA, MD: Twenty-eight degrees.
00:43:03
PEDRO J. del NIDO, MD: This is another secondary chord which needs to go. That actually -- I just felt what happened as soon as I cut it, and it's almost as if it was a tremendous amount of tension on the base of that leaflet. And as soon as I divided it, it just released. So it actually, I think that one will make quite a difference.
00:43:28
EMILE A. BACHA, MD: It's -- on the camera it looks like it's a big blob of tissue where you really can't differentiate anything from anything, but --
00:43:37

PEDRO J. del NIDO, MD: It's very thick, no question.
00:43:39
EMILE A. BACHA, MD: Right. But I'll say that when you see it through your loops, it's actually a little bit more easy to differentiate what's what, what's the secondary chordae, what's the primary chordae. Right now it looks like you're working on a piece of jello or something, so --
00:43:56
PEDRO J. del NIDO, MD: Not quite jello. It is -- in fact, unlike jello, it's really quite thick and quite hard to work with. Okay.
00:44:37
EMILE A. BACHA, MD: So this is going toward the LV outflow tract.
00:44:39
PEDRO J. del NIDO, MD: Yes. So this is another area that was quite thick. And the same issue here. I can still see -- but I'm beginning to see the back of the papillary muscles, which is what I need to see in order to mobilize those papillaries a little bit. And I can see down into the left ventricle, and can you just suction down there? So there are these attachments of the papillary to the free wall which need to be divided in order to actually get the papillaries to move more central in the ventricle. And that's what I'm doing right now. It doesn't affect the papillary at all. These are structures that shouldn't be there.
00:45:47
EMILE A. BACHA, MD: So those are attachments between the posterior free wall of the LV and the papillary muscle, the base of the papillary muscles.
00:45:52
PEDRO J. del NIDO, MD: Exactly. And they're on both sides. Today these are pretty prominent in this baby.
00:46:02
EMILE A. BACHA, MD: Yeah, we can see that very nicely here on the --
00:46:12
PEDRO J. del NIDO, MD: So I'm just -- and this will actually help with that posterior leaflet coaptation quite a bit as well. You can see that some of these can be quite rigid, quite thick, like this one. And that will definitely help. And this process is what --
00:46:32
EMILE A. BACHA, MD: In those patients that we've done that in we haven't seen much recurrence at all of that plastering of the posterior leaflet to the free wall of the LV.
00:46:42
PEDRO J. del NIDO, MD: No.
00:46:43
EMILE A. BACHA, MD: Once we've divided those chordae and released the papillary muscles, usually the mobility is markedly increased.
00:46:53
PEDRO J. del NIDO, MD: So I've done about as much mobilizing as I really need to do. Now I'm just going to reattach this.
00:47:01
EMILE A. BACHA, MD: Pedro, would you consider putting a pericardial patch here to build height?
00:47:06
PEDRO J. del NIDO, MD: Sure. You can. I don't think I need it today, but yes, you can. Most of what we're dealing with today is stenosis, so we're trying to improve mobility of the leaflets rather than dealing with regurgitation. If the problem were primarily regurgitation, then I think elongating a leaflet with a pericardial patch makes a lot of sense because that'll improve coaptation. So the way we reattach is just through a series of very fine interrupted sutures. I'm going to probably put about half a dozen of these in here to reattach it, and then we'll take a look at how the valve works.

00:47:47
EMILE A. BACHA, MD: Pedro, why wouldn't you run the sutures as opposed to interrupted?
00:47:51
PEDRO J. del NIDO, MD: The concern that I have about running is essentially the running suture will become almost like a purse string and lead to narrowing of the annulus. And if you're dealing with stenosis, anything that narrows the annulus is going to be counter to what you're trying to accomplish. So it takes a little bit longer, but I think in the long run it's better.
00:48:26
EMILE A. BACHA, MD: One question that's come from the audience is, "How long will the patient have to stay in the hospital?" Assuming everything goes well, which I assume it will, roughly 7 to 10 days.
00:48:38
PEDRO J. del NIDO, MD: Yeah, I think that's about right. We do know if you do quite a bit of EFE resection, those ventricles have some degree of diastolic dysfunction for a few days, and so they do require an extra day or two on the respirator. And you have to sort of plan for that.
00:48:55
EMILE A. BACHA, MD: Is there are lot of EFE that you can see in the ventricle?
00:48:59
PEDRO J. del NIDO, MD: Not really, no. I think most of the EFE is in the LV outflow tract, and I'm hoping to go back and address that through the aortic valve. I wanted to have a look at this mitral first, and then I'll go back.
00:49:19
EMILE A. BACHA, MD: Now at the first surgery, were you able to remove a lot of EFE? Sometimes in neonates with severe MS it's difficult to get into the ventricle and remove enough EFE.
00:49:29
PEDRO J. del NIDO, MD: Yeah, this child fortunately didn't have that much. I mean, he was actually a pretty good candidate for just -- I mean, he was essentially functioning as a two ventricle other than the LV outflow tract, which was quite obstructed. Can I have the 7-0, please?
00:50:05
EMILE A. BACHA, MD: So one of the structures one has to think about here is obviously the circumflex coronary artery. He's very much superficial to that, but any time you're --
00:50:16
PEDRO J. del NIDO, MD: And I'm also trying to stay on the ventricular side of that so that we don't have that issue. But yes, absolutely.
00:50:42
EMILE A. BACHA, MD: And that's 7-0 prolene you're using?
00:50:43
PEDRO J. del NIDO, MD: Yes.
00:50:44
EMILE A. BACHA, MD: And I think with those interrupted sutures, the tension is evenly divided on the -- one may say it's too small a suture to withstand ventricular pressures, but since you have evenly spaced interrupted sutures, and I think you're doing figure-of-eight sutures here.
00:51:03
PEDRO J. del NIDO, MD: Yeah, in fact, it's strong enough. And I would use 8-0 even in newborn babies. And this tissue -- the reason why I'm able to suture this tissue, which you really can't do in a normal valve, is simply because this is not normal leaflet. This is quite thickened valve tissue, and so that it actually -- the sutures hold quite well.
00:51:37

EMILE A. BACHA, MD: So it looks like scar tissue, but in fact we've sent in a whole bunch of specimens to be analyzed. And when you look at it, there's a lot of fibroblasts, a lot of myofibroblasts, and a lot of elastin and collagen, right, unlike scar tissue?
00:51:54
PEDRO J. del NIDO, MD: Yeah, it's very much almost like aortic wall, and so that it allows us to do this, whereas I wouldn't think about doing this in a normal mitral valve. We're almost done. I have a couple more to put in here just to do the final repairs or reattachments of this leaflet. Can I have an 8-0, please?
00:52:37
EMILE A. BACHA, MD: So what is the recovery process for this child? Again, as Dr. del Nido said, there can be a fair amount of diastolic dysfunction post-EFE resection. In this particular case here, we were not going to do much EFE resection per se and we're in fact improving his hemodynamic situation by relieving his aortic stenosis and subaortic stenosis. And this heart starts out with good systolic function, so I wouldn't expect that this child -- this child should only be better, and therefore his post-op recovery, I would expect it to be uncomplicated and not really have problems with diastolic dysfunction as opposed to a patient who is undergoing extensive EFE resection.
00:53:26
PEDRO J. del NIDO, MD: Yeah, there's no question that the EFE resection patients take a little bit longer on a respirator, unlike this child. This child should recover pretty quickly.
00:53:40
EMILE A. BACHA, MD: Obviously, another important parameter is what kind of ventricular function do you have going into the surgery. That's maybe the most important parameter. And again, this child had good systolic function and probably fairly good diastolic function as well. But if a patient's going in with poor ventricular function, then the recovery period may be a little bit more prolonged. On the other hand, we would say that if you have these lesions and you have poor ventricular function, then the need for surgery is greater even, and it is absolutely essential to repair any hemodynamic sequelae that you may have going in to the surgery.
00:54:25
PEDRO J. del NIDO, MD: Yeah, I think that we've found that to be an important point, and that is that you want to end up with as good a hemodynamic result as you can, and when you do that even fairly big-magnitude procedures are tolerated quite well.
00:54:40
EMILE A. BACHA, MD: Right. I'm going to answer a question here: "How did this patient's heart function differ from HLHS?" It's a gray zone really. There's no one box with HLHS and one other box non-HLHS. These patients are gradually HLHS -- there's a spectrum of HLHS. You can have a very, very small left ventricle and you would not be considered at all as a candidate for rehabilitation of left ventricle and you can have normal size LV with aortic stenosis mitral stenosis, and then there is a region in between where you have borderline LV size, and that's what we're talking about, the borderline left ventricle here.
00:55:19
PEDRO J. del NIDO, MD: Mm-hmm. Some cold irrigation, please.
00:55:23
EMILE A. BACHA, MD: And now we're going to test the mitral valve with saline injected into the left ventricular cavity. And he's keeping the aortic valve closed with his left index finger, where he can pressurize the left ventricle.
00:55:47
PEDRO J. del NIDO, MD: We're going to see some coaptation. I have to release these.
00:55:53
EMILE A. BACHA, MD: It's important to release the stay sutures because obviously they're putting up traction on the valve and you may have the false impression that there's a lot of

leakage, where it's only that you're hoisting up the valve. So you want to just lift the edge of the atrium just like that while exposing the valve. And now he can inject the saline.
00:56:33
PEDRO J. del NIDO, MD: Nerve hook, please.
00:56:44
EMILE A. BACHA, MD: That's the anterior leaflet of the mitral valve.
00:56:46
PEDRO J. del NIDO, MD: Yeah. And it has an area quite thickened right here. Knife. Can I have the knife?
00:57:26
EMILE A. BACHA, MD: So he's thinning out the atrial surface of anterior leaflet of the mitral valve, which the echo had shown this very nicely, the thickened edge.
00:57:47
PEDRO J. del NIDO, MD: You can see it really is quite, quite thick. Knife -- or scissors. That's a little bit better. Nerve hook, please. There was a ridge of tissue that was quite thick on the edge. Nerve hook. Okay, good. Malleable retractor again. So this is still also quite textile right here. And it's right on the edge of the anterior leaflet. And what we want to try to do is thin this out so that we can get better mobility. Knife, please.
00:59:32
EMILE A. BACHA, MD: That was nice. You can see how the plane is being developed and the normal tissue is falling away.
00:59:39
PEDRO J. del NIDO, MD: Chordal attachment is just below me here. But it's this [micsomatous] tissue that grows on these valves and becomes over time very thick and fibrous and really limits the excursion of the leaflet.
01:00:28
EMILE A. BACHA, MD: Do I have to wrap it up?
01:01:10
PEDRO J. del NIDO, MD: Okay, that's pretty good. Now let's look at the subaortic area again before we -- roll the table back towards me again, please.
01:02:00
EMILE A. BACHA, MD: So we've now changed the site of surgery, went back to a superior approach. And we're looking at the aortic valve and the subaortic area. So the aortic stay sutures have been picked up again. And Dr. del Nido is going to expose the left ventricular outflow tract.
01:02:33
PEDRO J. del NIDO, MD: And take a knife. So this is the subaortic membrane.
01:02:43
EMILE A. BACHA, MD: So here he's under the aortic valve and starting at the level of the right noncommissure on the septum away from conduction system. One of the issues here is to avoid injuring the conduction system. And he's moving toward the left of the septum towards the anterior leaflet of the mitral valve. And it's a continuum of this tissue growing, then I'm sure the same tissue is on the ventricular surface of the anterior leaflet of the mitral valve.
01:03:30
PEDRO J. del NIDO, MD: Unlike typical subaortic membrane tissue, this tissue is very adherent, just like the EFE. And you literally have to cut it away.
01:04:19
EMILE A. BACHA, MD: So Pedro, we're done with the webcast, so if you could -- if you have some final thoughts prior to --
01:04:29
PEDRO J. del NIDO, MD: Well, the only additional work that we need to do is I haven't addressed the secondary chords that we have looked at on the anterior leaflet of the mitral

valve. I think most of the subaortic membrane we've resected now, so that area looks pretty good. These are the secondary chords which we need to divide. Can I have a nerve hook, please? Then once we're done with that we'll close the aorta and then retest that mitral valve and see if we're happy with it and try to make sure that we've got a good, competent valve. I think the stenosis is pretty well addressed.
01:05:05
EMILE A. BACHA, MD: Yep. Thanks, Pedro. So now he's going for those secondary chordae that were impinging the movement of the anterior leaflet of the mitral valve as opposed to the posterior that he addressed by detaching the posterior leaflet. So as Dr. del Nido mentioned, there's still a little bit more work to be done, but essentially he's completed the operation. Those operations are difficult. You have to pay attention to detail, as usual in heart surgery. And one important feature is the interplay between the echocardiographer and the surgeon because once the procedure is done, we'll repeat the epicardial echo as a TE as well as a 3-D epicardial echo and make sure that we are happy with our repair. And should there be any residual lesions, we would not hesitate to go back on bypass and fix whatever residual lesion there is. We'll also typically measure intracavitary pressures to make sure we have not left gradient across the left ventricular outflow tract behind. So we'll measure LV pressure, ascending aortic pressure, left atrial pressure so we'll have a good assessment of whether we've done the job that we're supposed to do. I thank you for being with us, and bye-bye.
01:06:40
ANNOUNCER: This has been a live presentation of a complex congenital cardiac defect repair from Children's Hospital Boston. OR-Live makes it easy for you to learn more. Just click on the "request information" button on your webcast screen and open the door to informed medical care.
01:07:12
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