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Pulmonary Thromboendarterectomy for CTEPH

Announcer:
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Dr. Haft:
So, I'm going to talk about the surgical aspect. And maybe even before I do that, I'll just point out that that last slide that you showed about the impact of BPA on pulmonary endarterectomy, these two therapies are complementary; I don't think they're competitive. But I think having one in your program is going to enhance the other aspect of your program.

So, we've already learned about the European guidelines. And I just wanted to point out a couple of things here. Number one is, that if your patient has CTEPH, if they are operable, determined by an experienced center, the Class I recommendation is that they do undergo surgery. And they do point out that that assessment has to be done by an experienced pulmonary endarterectomy center and specifically an experienced pulmonary endarterectomy surgeon. And what does that mean, experience? I would suggest 100. It's a nice big round number. But I do think it takes a lot of experience to really be quite skilled and competent at the procedure.

If we're going to talk about the operation, we'd be remiss if we didn't mention Dr. Nina Braunwald. She's a cardiac surgeon from the 1960s and 70s. And you may notice that she is in fact a woman. And just being a cardiac surgeon in the 1960s as a woman is a big deal. But she was more than that. She was a leader among men. She was the chair of her department in San Diego. And she decided to tackle this phenomenon way back then. She partnered with Ken Moser, who I think we all recognize is likely the father of this particular disease, CTEPH. And using this sort of diagnostic imaging, which it's hard to imagine operating on somebody just based on these images. But she bravely took this patient to the operating room, and using thoracotomies and hilar collapse, she was able to remove this chronic organized thrombus and achieve a really good result in this particular patient. And this really opened up the possibility that these really sick and suffering patients could potentially be cured of this disease.

A lot of what we know now today about pulmonary endarterectomy I think we owe thanks to these two individuals. This is Stuart Jamieson, the surgeon that really defined and refined the operation that we do today, and this is Bill Auger, the pulmonologist who I think really made this into an everyday operation, the preoperative assessment, the perioperative care, and the postoperative management.

So from the surgical perspective, we see a patient that's being evaluated for pulmonary endarterectomy. So what am I thinking about? So, number one, does this patient have the disease? Do they have mismatch defects? And do they have functional limitations? Whether it's pulmonary hypertension or not, what's more important is are they symptomatic? And number two, is this idea of accessibility, how do we define accessibility? And we use the Jamieson classification based on the location of the obstruction. So this is a normal pulmonary angiogram on the right. And so Jamieson Class I would be at the branch PA level, which you can see from the arrow there. Jamieson II would be at the lobar level that you can see there, the interlobar trunk or the upper lobe trunk. Jamieson III is disease at the segmental level, and this is really obstructive disease where you see limitation in flow on the angiogram. And then Jamieson IV is that that subsegmental level. And I would say that any experienced pulmonary endarterectomy surgeon, Jamieson level III and above is surgical disease. For isolated Jamieson IV disease, it would really take the most experienced pulmonary endarterectomy surgeons to routinely

offer surgery to those patients.

And then again, as I'm evaluating these patients, the last thing we think about is the patients, are they a good surgical candidate? Their comorbid conditions and functional status, as Dan had suggested before in that 80-year-old woman with a lot of disease.

So the guiding principles of surgery. This disease is almost always bilateral just by nature. And so, it's always done through a median sternotomy so we can access both main pulmonary trunks. The operation is always done on cardiopulmonary bypass, and specifically using deep hypothermic circulatory arrest. And the endarterectomy always has to feather out into those distal tails, which is usually at the subsegmental level.

So we use standard cardiopulmonary bypass, nothing particularly interesting about that. We cool the patients again down to 18, which takes a long time. We do measure temperature in the nasopharynx, the blood, and in the bladder to make sure we have uniform cooling and it can take, you know, upwards of 1.5 hours to get those patients really nice and cold. We mobilize the right pulmonary artery off from behind the superior vena cava, you can see in that cartoon there. And we do the operation with a cross clamp and blood cardioplegia. Some centers don't, but we do and find it's additive. And then you make an incision in that right pulmonary artery, typically going beyond the take-off of that upper lobe branch which actually takes place within the pericardial space there. And then when we do our dissection, it's always done under hypothermic circulatory arrest.

This is the view from the surgeon's side. The surgeon stands on the left side of the table when they're doing the right lung, and then we switch sides when we do the opposite lung for better visualization. So you've got to find the right plane. This is part of the experience of doing the operation. This very simplistic cartoon sort of suggests it falls away, which it really doesn't, and being able to see that right plane and then tease it apart. You have to have the right instruments. This is the Jamieson suction device that you see here, very blunt, very fine-tip suction to get good visualization distally. And this was a game changer for us, these Wexler double-action, fine forceps that can get you into those very, very distal subsegmental branches. When you're in the right plane, it's pearly white, you know it when you're in that right spot. If you're a little bit too deep, it's pink because you peeled off the intima, and you're actually looking at the vasa vasorum that's shining through. And that's a problem that you have to recognize it and realize you're potentially in trouble if you're in that wrong plane. You start the plane wherever it's easy. Sometimes like in this cartoon, it's right where your incision is, and you can peel it off there. But oftentimes you have to start your plane out into those subsegmental branches. So wherever it is, is where you start. And then you continue that dissection distally using an eversion technique. So you're pulling the organized thrombus towards yourself, and pushing away that normal intima until it feathers out. And how hard to pull and how hard to push, again, comes with experience. So if you pull a little bit too hard, and it snaps, it will often retract out into those subsegments, and you may have lost it forever. So in that particular case that Dan showed, they probably had a specimen, it snapped back, and then they had to go and balloon it later. What's even worse though, is if you really put your foot on the bed and pull and you tear those distal arteries beyond where you can see, you may have a full-thickness injury and massive pulmonary hemorrhage, and you won't know it until about 2 hours later when you come off bypass. Again, the operation has to be done under hypothermic circulatory arrest for 20-minute intervals. No more than that. And then once you've done your dissection, and you've closed the pulmonary arteries, you re-warm the patient, this also takes a long time, it can be upwards of a couple of hours, particularly in patients who are large sized.

So here's a couple of videos here. This is going to get you inside that right pulmonary artery, before we've started the dissection – I'm just going to make you little dizzy here in the morning – and then back into that RPA. And you can see that wrinkly stuff there, that's nonobstructive organized thrombus, you can clearly see the obstruction at the segmental level. So this is Jamieson III disease there. But we're going to get a handle on that stuff that's in the - at the lobar level lifted and use it to do our eversion.

So I'll show you in the next picture here we've created that dissection plane and now we're doing the eversion technique. So we're going to grab that organized thrombus that we peeled off the wall, and then down into the segmental and even the subsegmental branches, you can see we're gently teasing away the organized thrombus from normal intima. I'm fighting with the camera right there, which is in that sort of confined space. But that's the technique of how you do it. And how hard you pull and how hard you push is something that really you gain with experience.

So what are the results of surgical treatment of this disease? So a lot of what we know comes from San Diego. So this is the experience, the early experience from Dr. Jamieson. This is a 20 plus-year-old paper with his first 150 cases. And you will see that the pulmonary artery pressures went from 80 down to the 40s. His early operative mortality was quite high, 9%. Remember, this is over 20 years ago. But what's notable in this publication is this is the circ arrest duration over time, and you can see not only have his circ arrest times gone down, but more importantly, or at least as importantly, the error bars are much tighter, just as you become more refined in doing the operation.

This is a more recent paper from the group in California, you can see now with thousands of cases under their belt. And we'll just show the results here. We'll skip here. And then the results there their pulmonary artery pressures, their PVRs are way down, the cardiac

output is down. Here you can see the PA systolic pressures down from 70s down to 40s, both in the early experience, and in their later experience. And their circ arrest times remain low in the 30s, despite adding a few extra surgeons to their group. These are their Kaplan-Meier survival, curves really good, long-term survival in this patient population. And in their more recent experience, operative mortality is now around 2%. So mortality has gone down substantially as they've gotten more refined on how they do the case.

Now you may say that's San Diego, how does that translate to the rest of the world? So here's the CTEPH registry in the U.S. This was about a dozen or so centers with passion in treating this disease surgically. These are World Heart Classification preop and then postop. You can see from World Heart Class III down to World Heart Class I and Class II. So, the vast majority of these patients have significant symptomatic improvement. And then if you look at other features, the orange is the – or the red is the patients who had surgery. This is use of oxygen cut in half, use of diuretics cutting in third, and use of PH-specific therapy cut in half. So really about half of these patients are effectively cured with a mortality around 4%.

We wanted to look at a wider swath. This is looking at the STS registry which captures basically 99% of all cardiac surgery procedures in the United States. And the database was available from 2012 on, for pulmonary endarterectomy. And this is the volume distribution of cases around the United States. And you can see, there's one center with 400 cases submitted over this time, San Diego. And then there's a handful of centers that are doing this with really consistent regularity. And then you got like a billion centers that did like one or two cases, right? So obviously, the question is, how do those patients do? And of course, they do much worse. Low-volume centers have higher complications, higher ECMO, higher reintubation, and higher mortality rates. So it's a volume to outcome relationship.

Now, this is our program in Michigan. And you can see that we've grown so we started slow, and now we're doing 25 to 30 cases a year, which we think is a healthy volume. And then you can see our PA pressures, systolic from 80 down to 40, mortality around 4%. Most of that is front loaded in our experience.

We take photos of the specimen like you saw in Dan's talk. And the purpose of it is not to have the big block M here on game day. But also, it's an important diagnostic tool for follow-up with these patients. So all these patients at 6 months, they get a regimented follow-up, 6-minute hall walk, echo, right heart cath, and a V/Q. And if those patients have evidence of residual obstructive pulmonary hypertension, then we'll look at that specimen. And then we'll look at the preoperative angiogram and decide should we pursue balloon pulmonary angioplasty. Now we give the photos of the specimens to the patients. And we have a member of our team who's a particularly talented artist, and she wanted to dress some of these up a little bit for the patients. And so, since this is some of her more recent work, this guy was a fireman. This guy liked to chop wood. This is a young lady who had a tattoo of a koi fish, and so we put that on our specimen.

So anyways, that's the nature of our program. I thank you very much for your time. There's my digits. If you guys have any questions in your own practices you want to talk about, feel free to reach out. And I think maybe we'll take it to the next speaker. Thank you very much.

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