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A Case of CTEPH: What To Do?

Dr. Channick:
Hello, my name is Richard Channick, I'm professor of Medicine at UCLA Medical Center, co-director of the Pulmonary Vascular Disease Program there. And today, I'm going to present a very interesting case of chronic thromboembolic pulmonary hypertension or CTEPH. And I'm going to ask you, what would you do in this case?

So we have a 56-year-old woman who was short of breath for a couple years. She did give the history of a pulmonary embolism four years prior and was also found to be positive for anticardiolipin antibody. There was no DVT found at that time. The patient was maintained on a oral anticoagulant and never gotten better. So she was still short of breath, wasn't seeing any real improvement in her exercise capacity. And more recently she started a note bilateral leg swelling, started on furosemide, diuretic. At that time, an ECHO was reported to show significant pulmonary hypertension and the patient was empirically started on sildenafil, but that led to no real change in any of her symptoms and so that was discontinued.

When we saw her in our clinic, these were her vital signs with her heart rate in eighties, good blood pressure, O two saturation, normal in 96%, no jugular venous distention, and a heart exam was significant for signs of pulmonary hypertension, along with a tricuspid regurgitation murmur and trace pedal edema.

So this is her x-ray. And I think what you can see here is pretty normal-looking lung fields, maybe a little hyperlucent and the heart looks a little bit large and looks like some right ventricular enlargement, but somewhat unremarkable chest x-ray. So because of the history, we got an echocardiogram and you can see here a little snippet of that clearly showing an enlarged, markedly enlarged right ventricle and right atrium with paradoxical motion of the septum and a reduced right ventricular function. The estimated right ventricular systolic pressure was 84. So very elevated. So this is what looks like significant pulmonary hypertension.

So we have a lady who has a history of a PE, persistent shortness of breath now for a few years, who has an ECHO, very suggestive of pulmonary hypertension. What would you do next? What would you order next? Would you get a V/Q scan, a CT angiogram, pulmonary function test, or a stress test? So think about which option you would pick. I think my opinion is that the best test to get next would be a V/Q scan. And the reason for that is that this patient, certainly, we have a high suspicion of CTEPH and really the best screening tests for CTEPH is a ventilation perfusion scan. And you can see here that scan. I can tell you that reality the patient actually had an outside facility, had had a CT pulmonary angiogram performed already and that was reported to show no pulmonary emboli, but as you can tell, and this is a very good example of the CT angiogram, missing chronic pulmonary emboli, cause this perfusion scan is not normal. And you can see they are really bilateral defects, perfusion defects. Some are smaller, more at the subsegmental, or small segmental level, but clearly an abnormal perfusion scan. So very suggestive of chronic thromboembolic disease.

The patient then went on to a definitive confirmatory test as per all protocols, which showed the following findings. And as you can see here, there are several abnormalities noted with bands across several upper lobe vessels, absent flow to the left upper lobe, diminished flow to both lower lobes with missing vessels. So these are some of the classic signs of CTEPH. And you can see with this slide the hemodynamics were quite abnormal and consistent with significant pulmonary arterial, pulmonary hypertension, I should say, with a mean of 52 and pulmonary vascular resistance as markedly elevated and a very reduced cardiac index of 1.3, right atrial pressure is elevated. So this patient's in right heart failure. I don't think anybody would argue that. And she has some signs of chronic

thromboembolic pulmonary hypertension. I should say that when we looked at this scan, this angiogram, I was a little bit worried. And the reason why I was worried is because those pressures are so high and that PVR are so high, but a fair number of the lesions look kind of distal, like they may be at the limits of accessibility. I did want to show you a lateral of view which was a little more reassuring and you can see on this lateral view, several of those lower lobe branches are just stopped, but there's also does appear to be some distal disease as well. So this is a borderline case I would say.

So what I would first ask you is is she a surgical candidate? Based on those hemodynamics and that angiogram and that history, would you offer this patient surgery, pulmonary endarterectomy? What about treating her medically with medical pH therapy in the preoperative setting? And what about alternative procedures like balloon pulmonary angioplasty? Those your options. So with that in mind, I just wanted to show you very quickly the treatment algorithm for CTEPH. So I think as we've alluded to patient has to be properly diagnosed with the disease. There's then an operability assessment. Is this disease accessible? Is it operable both from a point of view the anatomic location, as well as things like comorbidities? And if it's not operable, then there's a role for medical therapy and possibly balloon pulmonary angioplasty. So this is an algorithm that we go through all the time with patients as you're working up for CTEPH. So let me tell you what we did. As I mentioned, these lesions some were surgically accessible, but they're pretty borderline. So we offered her a pulmonary thromboendarterectomy, but gave her a higher estimated mortality. Severe pulmonary hypertension, very high PVR with borderline accessible disease, we gave her a higher mortality risk. So this was going to be a risky operation. She decided to go through surgery and had segmental disease removed. So no big surprises at surgery. What we call type three or level three disease was removed bilaterally. Postoperatively, we measure hemodynamics and her right atrial pressure was eight. Her pulmonary artery pressure was 64 over 22 that gave her a mean of 36, so not normal. Still somewhat elevated, but certainly reduced markedly from preoperatively. Notably, her cardiac output and cardiac index were significantly improved compared to what we saw preoperatively.

So that's where we're at, the patient recovered from surgery relatively and eventfully and was able to be discharged. But we have a patient who still has pulmonary hypertension despite getting a pulmonary thromboendarterectomy. And we need to ask ourselves, what more can we do for this patient? What more should we do for this patient? So my question to you is, would you offer this patient medical therapy postoperatively for persistent pulmonary hypertension? And if so, what medication would you use? What about balloon pulmonary angioplasty after surgery if there's still lesions present? Or would you just sort of sit tight and give the patient more time to recover, reassess, and let's say four to six months, before making a decision about use of medical therapy?

So what we did, again, make your decision, is we got the patient outta the hospital. She was feeling actually quite a bit better, probably due to the fact she was no longer in right heart failure. She was put on warfarin as we typically do for patients after endarterectomy. And in fact, her pre-discharge perfusion scan confirmed that there had been a benefit from the surgery. She had proof flow to both lower lobes and she had what we call stolen from the left upper lobes. So the flow left, left upper lobe to some degree and went to the newly open areas. Two months afterwards, we reassessed. So we didn't start anything immediately, reassessed at two months, she was better clearly than before surgery, but was not back to normal. She described significant exercise limitation that prevented her from doing the things she wanted to be doing. We repeated an ECHO at that time and you can see the results, mild to moderate RV enlargement, RVSP of 62 millimeters of mercury by estimate, and a TAPSE, mildly reduced to 1.6. We repeated a V/Q scan at that point to see whether any new defects or large defects and it really wasn't a change at all from her early postoperative scan. So no sign that she was reclotting, something we rarely see actually.

So I think we're still dealing with pulmonary hypertension. We confirmed that with the right heart cath that I'm showing you here, which showed pretty substantial precapillary pulmonary hypertension with a PA mean of 48, a wedge pressure of 10, and a cardiac index, mildly reduced to 2.3 that gave her a PVR of 8.2 wood unit. So, she still has significant pulmonary hypertension, still symptom limited.

So what would do you do? Would you send her for a balloon angioplasty say, well, let's see if there are any lesions to open up and open them up? Would you say, well, this is the best we're going to do and you're going to have to live with it? Or would you start the patient on targeted medical therapy for residual pulmonary hypertension? Those are options. Those were our options. What did we do? So our thought process was based on residual pulmonary hypertension, no evidence for new pulmonary emboli. And we felt comfortable that the surgeon had gotten out what was there. We decided to treat this patient with medical therapy, targeted medical therapy for persistent pulmonary hypertension after endarterectomy. Now, I should say that some of these patients, we may be able to do a pulmonary angioplasty on residual lesions. Although, I do have to acknowledge that that's still in the learning phase and doing balloon pulmonary angioplasty after a pulmonary endarterectomy is not being done that frequently. And you really would want to convince yourself that they're discreet lesions. But I think the role of medical therapy in these patients is pretty clear and pretty well-proven. And in fact, in this particular patient, she got better. So she improved her clinical status, functional class improved from a three to a two, her ECHO showed improvement in the right particular function, still not normal, still not normal, but we're considering additional medications for this patient, for her residual pulmonary hypertension.

But I think this is a good example of a patient who was a high-risk case based on the preoperative relationship between the hemodynamic severity and the radiographic burden of disease, severe pulmonary hypertension and borderline accessible disease. The assessment of operability remains a subjective exercise. I have to acknowledge. Clearly should be done by those experienced in evaluating and making those decisions, a true CTEPH team. The role of bridging medical therapy in high-risk patients which we alluded to and considered, really hasn't been proven, needs careful investigation and certainly should not delay surgical intervention. And there's some data that it may in fact delay. So be very careful about trying medical therapy before surgery, no clear proof that that's beneficial. And I think maybe most importantly, careful follow-up assessment in a patient like this after surgery is really critical, because a significant number of these patients may have some residual pulmonary hypertension. A patient like this we could almost predict she was going to have some given how borderline accessible her disease was and really have a low threshold for treating a patient, for evaluating a patient, for residual pulmonary hypertension, and giving them appropriate medical therapy as we did in this particular patient. So those are your take on messages, and I hope you found this case interesting. Thank you for your attention.