

- ROBERT FRANTZ:** Hi there. I'm Dr. Robert Frantz, director of the Mayo pulmonary hypertension clinic, and I'm pleased to be here today accompanied by my good friend and colleague, Dr. Gurpreet Sandhu. Dr. Sandhu is the director of the cardiac catheterization laboratory, and an experienced interventional cardiologist. Gury, thanks for being here today.
- GURPREET SANDHU:** Thank you, Bob.
- ROBERT FRANTZ:** So we're here to talk about a condition called chronic thromboembolic pulmonary hypertension. Now that's a real mouthful, but what it really means is that sometimes when people have blood clots, they fail to clear them out of the lungs and are left with residual obstruction of the blood vessels into the lungs. This can cause significant shortness of breath and exercise limitation.
- GURPREET SANDHU:** So Bob, do want to talk about where these clots come from and what their natural history is?
- ROBERT FRANTZ:** Well, any of us are at some risk for forming blood clots, particularly in our leg veins, for example, after a long car ride or working under a tractor for an afternoon as the farmers in the midwest are wont to do. And sometimes those clots travel into the lungs and obstruct them. Now the standard treatment for that is blood thinners in order to dissolve the clots, but sometimes instead of dissolving the clots the clots actually become organized and hard like a scar and obstruct the lung vasculature.
- GURPREET SANDHU:** So that reminds me of a young patient we saw a couple of years ago, a 33-year-old gentleman, very active individual. So he had a history of blood clots. And then he came to see you in clinic with this progressive shortness of breath. So what did you do with him?
- ROBERT FRANTZ:** Well he's a really interesting gentleman who likes to actually hike through the mountains at fairly high altitude at fast pace. And so he was in quite good physical shape. And in fact, when we did exercise testing, he had quite well preserved exercise capacity, and didn't have any resting pulmonary hypertension, that is, his pressures in the lung vessels were OK when he wasn't doing anything. But he complained quite bitterly of not being able any longer to do the kind of high altitude sojourning that he liked to do. And so we went ahead with imaging in order to look at the lung vasculature in terms of whether the flow into the lungs was the way it should be.
- GURPREET SANDHU:** So on the top two images, basically the one on the left side shows the perfusion or the air entering into the lungs. That looks quite normal. The image on the top right, the lower segment there shows a lack of blood supply. So the image on the left and right should be pretty much similar, but the one of the right has something missing from the lower part of the left lung.
- ROBERT FRANTZ:** So essentially, there should be a good match between where the air goes and where the blood goes, because that's the point of the lungs, to exchange oxygen. And if you have air going a place where there's not good blood flow, that's sort of wasted ventilation. You're breathing in and out but that air is doing exactly nothing. And that makes you feel out of breath.
- GURPREET SANDHU:** So is there a better way of trying to identify where the arteries are, where the lung volume is as opposed to this black and white image?

ROBERT Well there really is a better way. So we've started to do this more volumetric approach where we use a short acting isotope and use CT imaging and look at the lungs. And in this next image, you can see this whole area. So **FRANTZ:** this is looking at this young man from the front. And you can see in the left lower area there is very little of the green, that is the isotope, going in that area where there should be good flow. And indeed we can overlay the c.t. Images that are from this chest CT angiogram demonstrating that there is obstruction to flow pretty much that whole left lower lobe segment.

GURPREET So here the right lung looks pretty much normal, and the left lower lung has lack of blood supply. So what would **SANDHU:** you do in a case like this? What is a normal, traditional way of treating these? Well the tradition would be to do a median sternotomy, open the chest, and shell out that material, and then close the chest again. Now that's a big operation. And it could be done with relative safety, but for a young man who says, you know, actually, you're telling me I don't have any high lung pressures at rest, I have pretty good exercise capacity, and you want to open my chest? I don't think so.

So we're talking a little bit today about an evolving strategy, which is balloon pulmonary angioplasty, using balloons to open up blood vessels in the heart, in the lungs, which we would sort of extrapolate from what we do in the heart circulation, which you have an artery narrowing, and do that in the lungs.

GURPREET And normally if someone had larger clots sort of in the central arteries, the larger arteries, then it would make **SANDHU:** sense to have a surgeon open up the chest and remove all the clots from both lungs. Here we have just one area of one lung, and there again, opening up the entire chest doesn't always make sense.

ROBERT Right. So you can see on the right of this image there is sort of an overlay of the angiogram showing this area in **FRANTZ:** the left lower lung that does not have proper blood flow, and then we can do this procedure where we inject contrast selectively into that segment with a catheter put through the vein in the leg. So the next step for this kind of a problem is to take even a more fine picture of the area of interest by doing a selective pulmonary angiogram into that segment. If you look at this picture, you can see that there is an injection into the left lower lung zone where we showed you those defects on the perfusion scan. And what do we see here, Gury?

GURPREET Normally what you would expect to see here is much larger blood vessels and a lot more blood rushing down. So **SANDHU:** this should essentially turn into a shade of gray as a blood fills all the little branches, but here we aren't seeing a very good blood supply.

ROBERT Right. And there are actually multiple branches here, and so in order to do this safely, you ended up using three **FRANTZ:** different wires down these vessels with graded balloons down each of them, and a bigger balloon at the beginning part of them as shown here. So there's the three little wires going down the little branches to protect them, and then a bigger balloon up top. And with this, we sort of do serial balloon dilatations the stations until we feel we have a good angiographic result.

GURPREET And now we see the beginning of the procedure on the left side, and then after we had opened up blockages in **SANDHU:** three branches we see much better blood flow in the image on the top right. So everything looks a little larger, a little more brisk flow. And this is what we would consider a fairly good result in terms of improving circulation to that part of the lung.

ROBERT FRANTZ: Right. And the other way we can follow this up is to do our perfusion imaging again with the isotope and see if we've actually restored good flow into that segment by that technique. So on the top here, you can see these images showing this defect in that left lower lung zone before we did the pulmonary balloon angioplasty, and then afterwards you can see that there's more perfusion into that segment as you can see down below. And this correlated with this patient feeling better, going back to high altitude trekking, and really able to resume these activities that he loves to do.

And so in fact, he called me recently and said that he wants to go to Everest base camp this autumn. Is it OK to do that? And I kind of scratched my head for a few minutes, I said, yeah, you can do that. There's no reason you can't do that. He'll stay on his blood thinners to keep him from having more blood clots, and so this was an extremely gratifying result.

GURPREET SANDHU: This is a good thing to remember, that the majority of patients who have blood clots, about 95% or more completely go away with blood thinners, but there are somewhere in the range of 1% to 5% who never really resolve the blood clot. They still keep having symptoms. So for those patients there are definitely options that can be used to make them feel better.

ROBERT FRANTZ: So the key here is if you've had blood clots then you really should have follow up imaging done six months later after you've been on blood thinners for an adequate period of time to make sure these clots have fully resolved. And if they have not and if you're having symptoms of some exercise tolerance or shortness of breath, you may have this condition called chronic thromboembolic pulmonary hypertension

Gury, it's been great to talk to you about this rare problem today. It's not maybe as rare as we used to think. I saw a recent publication that suggested if people who've had blood clots and routinely have follow up perfusion lung imaging done six months later, that up to 25% may have some residual perfusion defect. So it actually may be more common than we realized.

GURPREET SANDHU: This was great talking to you, Bob. Thank you.

ROBERT FRANTZ: Well, we hope we taught you something about chronic thromboembolic pulmonary hypertension today, and Mayo expertise in dealing with this difficult problem. Thanks for your attention.