

November is PH Awareness Month and CTEPH Awareness Day

Could you have CTEPH? Find out with a V/Q scan

If you have pulmonary hypertension (PH), make sure you get tested for chronic thromboembolic pulmonary hypertension (CTEPH). CTEPH is a form of PH caused by chronic blood clots in the lungs.

The first step to identify or rule out CTEPH is a ventilation/perfusion (V/Q) scan.

A V/Q scan is a radiologic test that screens for chronic or undissolved blood clots in the lungs. The clots can lead to chronic thromboembolic pulmonary hypertension (CTEPH).

A V/Q scan takes two images of your lungs and compares them to each other: One identifies ventilation (air flow); the other, perfusion (blood flow). Both scans use nuclear imaging to take pictures of your lungs.

During the ventilation scan, you will breathe in a small amount of safe radioactive gas through a breathing mask or mouthpiece. Pictures from the scan can show

areas of the lungs that don't receive receiving enough or retain too much air.

The perfusion scan shows where blood flows in the lungs. To check blood flow in the lungs, you will receive an injection of a safe radioactive dye, called a tracer, through an IV inserted in your arm. Pictures from the scan can show areas of the lungs that don't receive blood as expected.

If the lungs are working normally, blood flow on a perfusion scan exactly matches air flow on the ventilation scan. Areas that are ventilated but not getting blood flow (on the perfusion scan) are called mismatches and raise strong suspicion of blood clots.

If your test is abnormal, you might need additional testing, such as a pulmonary angiogram. Your PH care team will be notified and will contact you with further instructions or treatment options.

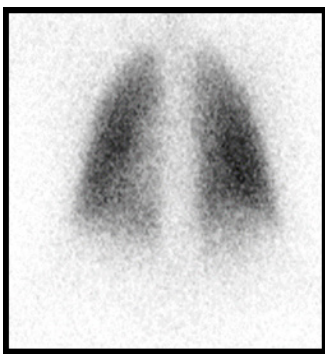


Figure 1 shows normal air flow in healthy lungs.

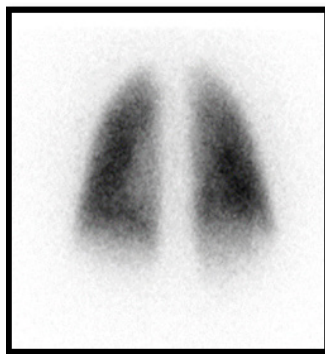


Figure 2 shows normal blood flow in healthy lungs.

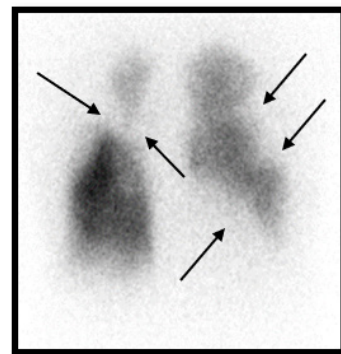


Figure 3 shows white areas (indicated by arrows above) where blood clots are preventing normal blood flow through the lungs.



Images courtesy of the Pulmonary Vascular Research Institute

Patient Story:

Noël Holly



I was diagnosed with chronic thromboembolic pulmonary hypertension (CTEPH) in 2008 after being hospitalized with massive blood clots in both of my lungs.

A year before, I had been coughing nonstop and was severely out of breath with minimum exertion. I was admitted to the emergency room in November 2007 after a deep vein thrombosis (DVT) in my leg traveled to my heart and into my lungs. It was a silent clot. I never had common DVT symptoms, such as redness, bruising, swelling or pain. While in the hospital, I had an ultrasound of my legs, a CT scan of my lungs and a pulmonary function test.

After 10 days in the hospital, I went back to work. I was put onto an oral blood thinner and referred to my doctor for follow up visits. In that time, the coughing worsened, and all other normal life functions were almost impossible. The doctor ordered an echocardiogram and determined that I had pulmonary hypertension (PH).

A specialist at the University of California-Los Angeles conducted more tests, including a V/Q scan. He showed me the test results and explained that my PH was caused by blood clots, also known as CTEPH. He told me my

clots were inoperable because they had become too fibrous.

I transferred my care to a specialist near me. After we discussed my case and treatment options, he started me on Tracleer. While it took some time, I noticed I could increase my activity without feeling like I could hardly breathe. In time, my doctor added two more therapies, Adempas and Uptravi. These three medications and lifestyle changes have helped me to continue to function and enjoy life.

After my diagnosis, I went on disability. I made the decision to take a step back and let go of several obligations. I accepted that no matter how much effort I put into something, it wouldn't be enough. I stopped trying to "do it all" like I used to before my diagnosis.

Even though CTEPH has been a life-changing diagnosis, I'm so thankful for my doctor and the people I have met in the PH community who understand and support me. I'm a former PHA support group leader, and now I give back to this community by volunteering for the PHA Support Line. A PH diagnosis can be frightening and overwhelming, and it's important to me to be that support for others.

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