

Transcript Details

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When to Suspect ATTR-CM: Clues for Earlier Diagnosis

Announcer:

Welcome to *Heart Matters* on ReachMD. On this episode, we'll hear from Dr. Ronald Witteles, Professor of Cardiovascular Medicine at Stanford Medicine and Co-Director of the Stanford Amyloid Center and Multidisciplinary Sarcoidosis Program. He'll be sharing clinical clues that may signal transthyretin amyloid cardiomyopathy, or ATTR-CM. Here's Dr. Witteles now.

Dr. Witteles:

So the question of how to suspect the disease—I think about it in two ways. There's looking for some of the extracardiac red flags, and then there's, well, what should you look for in the heart? And do you even need the extracardiac red flags?

In terms of extracardiac red flags, there absolutely are some, and the most common by far are what are collectively termed the “orthopedic manifestations” of the disease. Wild-type, at least for ATTR amyloid, most likes to go to two places: the heart, of course, and ligaments and tendons. And the ligament and tendon involvement will cause things like carpal tunnel syndrome, trigger finger, biceps tendon rupture, rotator cuff pathology, and lumbar spinal stenosis. All of these are clearly associated with the disease. If you look for it, you can often find ATTR amyloid in the ligaments and tendons that causes this pathology. But of course, many of these things, like carpal tunnel, are pretty common. And realistically, if a cardiologist is asking about it, you've already probably won the battle, meaning that they're already thinking about the disease. Otherwise, it's not that common for cardiologists to pay attention to these orthopedic manifestations, and I understand there's a lot for cardiologists to keep track of. But it is worth remembering, and certainly part of the patient education, because patients know if they've had carpal tunnel, or rotator cuff pathology, et cetera. So if you see that clustering with any of the cardiac manifestations, you really should think about the disease.

But the second part is to say, you don't need extra cardiac manifestations, and actually you don't need much to trigger the testing for this disease. So what is needed? Number one is the right demographics. So this is a disease that tends to be slowly progressive, and therefore it manifests in people who are older, most commonly 70s and 80s. Now, you can see it in people in their 60s, and occasionally somebody who won't present until their 90s. But it would be extremely uncommon for at least the non-inherited wild-type form of the disease to strike people in their 50s.

Now, there is a smaller number of people with inherited forms of the disease that can strike earlier. In the United States, the most common mutation is called V122I or V142I. And this mutation is present in over three percent of Americans of Sub-Saharan African descent. And so if you are caring for a patient of Sub-Saharan African descent, keep in mind that, A, it's a little more likely that they will have this disease, and B, it can present a bit earlier. So let's say mid-to-late 50s wouldn't be uncommon. So that's one part of the demographics.

The other part of the demographics is that it is more common in men, and we don't understand why. I think there's a lot of proper concern to say, “Is it underdiagnosed in women?” I'm sure the answer to that is yes. But it's also true unequivocally that it is far more common in men. And so I think it is reasonable to say, “I'm going to particularly think about this disease in older, let's say, 60, 70, 80-year-old men. Not that I'll ignore it in others, but that's who I'm going to really have front of mind.” And then that plus a thick heart, meaning, of course, for cardiologists what we'll often call LVH, even though in this disease it's not actual hypertrophy; it's increased wall thickness because of amyloid deposits. But if you have somebody who's the right demographics and they have a thick heart on echocardiography, that honestly should be enough to trigger the evaluation, particularly because, as we discussed earlier, it's not difficult to do the evaluation—simply monoclonal protein testing and a PYP scan.

Announcer:

That was Dr. Ronald Witteles discussing clues that can help us recognize ATTR-CM. To access this and other episodes in our series, visit *Heart Matters* on ReachMD.com, where you can Be Part of the Knowledge. Thanks for listening!