Transcript Details

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Complexities in ATTR-CM Care: Key Considerations for Accurate Diagnosis

Announcer:
You're listening to *Heart Matters* on ReachMD. On this episode, we'll hear from Dr. Gautam Nayak, a cardiologist at Kittitas Valley Healthcare in Ellensburg, Washington. He'll be discussing the current diagnostic approach for patients with suspected transthyretin amyloid cardiomyopathy, or ATTR-CM. Here's Dr. Nayak now.

Dr. Nayak:
So our current diagnostic and management approach is pretty involved. The first thing is you just have a clinical suspicion for ATTR, and we'll break it down into cardiac and noncardiac findings. So in terms of cardiac findings, it's heart failure symptoms first and foremost. Patients will present with dyspnea, lower extremity edema, and in the more extreme conditions, things like hepatic congestion and ascites. You will occasionally see patients come in with syncope, an advanced AV block. Infiltrative conditions like ATTR affect the conduction system, and pacemaker implantation is often required for these patients. Now, you do have to be careful because there's overlap with autonomic dysfunction, which is also seen in these patients, and that can also cause syncope.

Atrial fibrillation is very common in these patients, and oftentimes they'll actually present with not faster, but slower atrial fibrillation as their initial presentation. These patients have a higher risk for thromboembolism due to the advanced atrial myopathy that's associated with ATTR, so you have to have a really low threshold to anticoagulate these patients. And in fact, even despite anticoagulation, you can see evidence of left atrial appendage thrombus, so I typically will do a transesophageal echo before I cardiovert these patients because even on anticoagulation, you can still see thrombus.

The rhythm control when ATTR patients have AFib is really difficult, so amiodarone is commonly used, but we oftentimes will just focus on rate control. Ventricular tachyarrhythmias can occur in these patients, but they're far less frequent, and ATTR amyloidosis and aortic stenosis coexist. There's been studies that have suggested that up to 16 percent of patients with aortic stenosis undergoing TAVR, or transcatheter aortic valve replacement, for severe aortic stenosis have ATTR amyloidosis, so they do coexist, and they're commonly seen together.

Now, in terms of noncardiac findings, there's a few things to really keep in mind. Patients with ATTR amyloidosis will oftentimes present with fairly marked orthostasis or postural hypotension and dysautonomia. They also get peripheral nerve disease, and it's usually an axonal neuropathy. Axonal neuropathies present with progressive muscle weakness, especially in the feet and hands, accompanied by sensory symptoms like numbness, tingling, burning pain. And it usually is something that can lead to gait instability from the muscle weakness as well as the pain. And then you'll see the autonomic dysfunction not only occur in terms of their posture and postural hypertension but also in the bowels as well as other nerve fibers, where you get autonomic innervation.

The diagnostic approach with labs is really kind of critical. You want to rule out AL amyloidosis first, and we typically do that with lab tests to look for free light chains as well as a serum protein immunofixation and a urine protein immunofixation. In terms of diagnosing ATTR cardiomyopathy, you typically will start with things like an EKG and an echo. The hallmark EKG finding is a discordance between LV wall thickness on, say, the echo and the QRS voltage, which can be reduced, but I would argue that oftentimes what I will see is actually normal voltage on the EKG with left ventricular hypertrophy on the echo, and so that should also clue you in to think about something like ATTR. Additional findings on the echo are dilated atria, interatrial septal and valve thickening, and pretty marked diastolic dysfunction and resultant elevations in the pulmonary pressures. If you have the ability to do strain imaging, you'll see this characteristic bullseye or apical sparing pattern on the global longitudinal strain imaging, and so that is oftentimes very helpful and can be fairly specific for amyloid.

Once you have that clinical suspicion, you've got an EKG and an echo that's making you think about ATTR, and you've ruled out AL, you want to then, if you can, do bone tracer cardiac scintigraphy or a nuclear scan. We typically use technetium pyrophosphate as the tracer for this. It doesn't take very long. It's about a 15- or 20-minute scan. But there's actually been increasing global shortages of pyrophosphate, so in that case, you can maybe use an alternate tracer like HMDP. If you don't have access to a nuclear scanner but can get in a cardiac MRI, that's also very reasonable. It's not specific for ATTR versus AL amyloidosis, but you can see evidence of infiltration, and if your clinical suspicion is high, that should give you a pretty good clue as to what's going on, especially if you've already ruled out AL with labs. And then, of course, there is always a role for endomyocardial biopsy or biopsy of other locations, but it's becoming less and less common.

Announcer:
That was Dr. Gautam Nayak talking about diagnosing transthyretin amyloid cardiomyopathy. 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!