Heart House Roundtables

American College of Cardiology

Top 10 Takeaways:

The ACC's Heart House Roundtable, *Cardiac Amyloidosis: Clinical Gaps and Unmet Needs*, identified the following 10 key opportunities when it comes to caring for patients with cardiac amyloidosis:

01 IDENTIFY CLINICAL CLUES EVIDENT ACROSS DIFFERENT DISCIPLINES IN MEDICINE

Recognition of the non-cardiac (orthopedic, neurologic, renal, and gastrointestinal) manifestations of systemic amyloidosis is essential to facilitate early and accurate diagnosis. Educational initiatives that reach specialists of a variety of different clinical disciplines will be important to raise the index of suspicion and trigger appropriate steps in the diagnostic pathway.

02 AVOID MISTAKES IN NON-BIOPSY, IMAGING-BASED DIAGNOSIS

The principal errors in the diagnosis of transthyretin cardiac amyloidosis (ATTR) involve the inappropriate application of bone avid tracer cardiac scintigraphy without testing for a plasma cell disorder, failure to accurately interpret plasma cell disorder testing in the clinical context, or failure to proceed to invasive cardiac biopsy when appropriate. In addition, bone avid tracer cardiac scintigraphy must be performed with single photon emission computed tomography (SPECT) and ideally SPECT/computed tomography (CT) imaging to distinguish blood pool activity from myocardial activity and avoid false-positive scans as has been observed with planar imaging only.

03 DON'T FORGET ABOUT GENETIC TESTING

A. Genetic testing for diagnosis in patients with known ATTR

All patients with ATTR should undergo genetic testing to assess for variant versus wild-type disease regardless of age as there are implications for first-degree relatives (autosomal dominant inheritance). In addition, some treatments currently available (in 2021) are indicated only for those with variant disease.

B. Genetic cascade testing and population screening

First-degree relatives of patients with hereditary amyloid transthyretin amyloidosis require genetic counseling regarding the many ramifications of undergoing genetic cascade testing. The benefit of more widespread screening of high-risk populations may prove useful in the future, though unpredictable penetrance of known pathogenic variants make genetic counseling challenging.

04 ENSURING THE RIGHT TEST IN THE RIGHT PATIENT: WHOM TO SELECT FOR ACTIVE ASCERTAINMENT

Active ascertainment (screening) would allow for early diagnosis of ATTR and will be best utilized in cohorts with high pre-testing probability, such as those with increased wall thickness and a non-dilated left ventricular cavity. However, care must be taken when generalizing diagnostic tests validated in high pre-test likelihood populations to low pre-test likelihood populations. In addition, the cost-effectiveness of screening high-risk populations must be established.

05 PROMOTE ONGOING STUDIES OF EXISTING AND NEW THERAPEUTICS IN PATIENTS WITH TRANSTHYRETIN CARDIOMYOPATHY

Studies will offer insight into the impact of new therapeutics on cardiovascular endpoints as well as the role of TTR silencers versus stabilizers in disease management.

06 IMPROVE IDENTIFICATION OF MARKERS OF RESPONSE TO THERAPY

Prospective multicenter collaborations are essential to define the role of biomarker and imaging surveillance in patients with ATTR receiving therapy to determine treatment efficacy.

07 DEVELOP GUIDANCE FOR ADVANCED HEART FAILURE (HF) MANAGEMENT

Further guidance is needed for advanced HF transplant management. The indications and contraindications for heart transplantation in patients with cardiac amyloidosis are not clear and vary by center. Though most consider symptomatic gastrointestinal involvement and autonomic dysfunction, as indicated by orthostatic hypotension, contraindications to transplantation. Guidance formulating the listing criteria would be useful.

08 EXPLORE OPPORTUNITIES TO ESTABLISH AMYLOIDOSIS CLINICS AND ACCREDITATION CRITERIA

Given the multisystem impact of cardiac amyloidosis, multidisciplinary amyloidosis clinics comprising specialists from cardiology, neurology, orthopedic surgery, hematology/oncology, gastroenterology, nephrology, as well as pharmacists and advanced practice providers would improve the evaluation and management of these patients. Access to appropriately skilled pathology and cardiac imaging support is essential. Accreditation criteria or competency designations would be helpful to facilitate patients navigating to qualified centers for care and establish networks of high-quality programs that can collaborate.

09 EXPLORE ALTERNATIVE MODELS OF PAYMENT

The care of the amyloidosis patient is challenging owing to clinical complexity and time required for optimal care. These features are not favorable in a fee-for-service payment model that emphasizes volume. Alternative payment models that explore bundled or value-based compensation may better align with actual care delivery and support the development of clinical programs in non-tertiary care centers.

10 ADDRESS DISPARITIES OF CARE

Cardiac amyloidosis care is frequently administered at select centers of excellence thereby creating a disparate system of access. Telehealth has the potential to overcome disparities by permitting consultations from experienced providers. Federal and state oversight agencies can assist by removing barriers to permit billable interstate telehealth referrals.