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Screening for transthyretin amyloid cardiomyopathy in everyday practice

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Background: Transthyretin amyloid cardiomyopathy (ATTR-CM) is a life-threatening, progressive, infiltrative disease caused by the deposition of transthyretin amyloid fibrils in the myocardium. ATTR-CM is often overlooked as a cause of common cardiovascular conditions in older heart failure patients. Lack of disease awareness, misdiagnosis and misconceptions about the diagnostic process are, in part, responsible for a delay to diagnosis and application of appropriate patient management strategies, which can lead to a poor prognosis. Today, with the availability of bone scintigraphy as an inexpensive, non-invasive tool to diagnose ATTR-CM, the rationale to screen for this disease within certain populations of patients is increasingly warranted.

Purpose: Here, we aimed to develop a clinical framework to aid clinicians in identifying appropriate patients to be screened for ATTR-CM among the wider heart failure population.

Methods: An international panel of 11 amyloidosis experts convened to develop a consensus on the patient characteristics and clinical scenarios that further heighten suspicion or support the possibility of an underlying ATTR-CM, as well as a list of ‘red flags’ that should provide further evidence to support screening for ATTR-CM in general practice.

Results: The expert panel advised that men over 65 years or women over 70 years with heart failure and left ventricular wall thickness ≥14 mm should be screened for ATTR-CM. Some of the cardiac ‘red flags’, providing further evidence as to the possibility of ATTR-CM as an underlying condition, include: reduction in longitudinal strain with apical sparing; discrepancy between left ventricular wall thickness and QRS voltages; the presence of atrioventricular block and infiltrative features; and a diffuse late gadolinium enhancement with cardiac magnetic resonance. The presence of extra-cardiac symptoms such as sensorimotor polyneuropathy, dysautonomia and a history of bilateral carpal tunnel syndrome further support the possibility of ATTR-CM. Once a suspicion of ATTR-CM has been raised, a non-invasive diagnostic approach is recommended. The combination of bone scintigraphy and free light chain testing is the basis for the appropriate non-invasive screening of patients with suspected ATTR-CM.

Conclusion: This framework should assist clinicians in recognising patients at risk of having an underlying ATTR-CM. Awareness and timely diagnosis of ATTR-CM will facilitate the provision of optimal patient care.
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