CARDIAC AMYLOIDOSIS

Cardiac amyloidosis is an under-recognized and potentially fatal cause of heart failure and other cardiovascular manifestations. It is caused by deposition of misfolded precursor proteins as fibrillary amyloid deposits in cardiac tissues.

Most common subtypes of cardiac amyloidosis

Light chain amyloidosis (AL) – Hematologic malignancy with multisystem amyloid deposition, most often involving heart and kidney.

Transthyretin amyloidosis (ATTR) – Misfolding of the hepatic protein transthyretin due to either a gene mutation (hereditary type, hATTR), or ageing (wild-type, wtATTR), with predominantly heart and peripheral nerve involvement.

Epidemiology

Cardiac amyloidosis **is more common in older patients, and more common in men** than women.

AL amyloidosis has **an approximate annual incidence of 10/million**.

hATTR is endemic in certain regions but **relatively rare in Canada.**

wtATTR is likely underdiagnosed in older patients with heart failure and **may account for over 10% of HFpEF cases in this demographic**.

RED-FLAG SYMPTOMS

New onset heart failure + one or more of the following:

- Unexplained increase in LV wall thickness
- Low-flow low-gradient aortic stenosis with preserved LVEF (in patients >60 years old)
- Carpal tunnel syndrome (bilateral)
- Established AL or ATTR in non-cardiac organ/system
- Peripheral sensorimotor neuropathy and/or dysautonomia

How do you investigate suspected cardiac amyloidosis?

Standard HF workup: cardiac imaging (ECHO, CMR), troponin and BNP/NTproBNP

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Screen for plasma cell dyscrasia: serum and urine protein electrophoresis/immunofixation, serum free light chain assay

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If AL suspected – Hematology referral, tissue biopsy

If ATTR suspected (AL screen negative) – Tc99m-PYP scan (if unavailable, cardiac biopsy)

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Genetic testing if ATTR confirmed, to differentiate hATTR from wtATTR

Management

Cardiac Sequelae

1. Diuresis, sodium restriction for heart failure
2. Cautious use of β-blockers/ACEI/ARBs
3. Anticoagulation for atrial arrhythmias
4. Pacemaker for symptomatic bradycardia

Disease Modifying Therapy

1. AL – chemotherapy +/- stem cell transplant
2. wtATTR or hATTR with NYHA class I-III heart failure – Tafamidis
3. hATTR with polyneuropathy – Inotersen or patisiran

To learn more about Cardiac Amyloidosis please visit us at **CCS.CA**

Canadian Cardiovascular Society
Leadership. Knowledge. Community.

Société canadienne de cardiologie
Communauté. Connaissances. Leadership.

This infographic is an informative knowledge translation tool that features information based on the Canadian Cardiovascular Society/Canadian Heart Failure Society Joint Position Statement on the Evaluation and Management of Patients with Cardiac Amyloidosis. March 2020.