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#MADEEASY

AMYLOIDOSIS

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What is AMYLOIDOSIS?

Amyloidosis constitutes a group of disorders characterized by extracellular accumulation of amyloid fibrils, leading to organomegaly and organ dysfunction.

Pathogenesis

- ***Overaccumulation of misfolded proteins aggregate to form non-branching amyloid fibrils.***
- ***These amyloid fibrils are deposited in extracellular space.***

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Amyloid Precursor Proteins

AL (Amyloid Light chain)

- ***Derived from the light chain of immunoglobulin.***
- ***Associated with Primary amyloidosis.***

Tips to remember: ***You can imagine AL amyloid as Aloo, it is like the most primary and light to eat as well.***

AA (Amyloid Associated protein)

- ***Derived from Serum Amyloid A.***
- ***Associated with chronic inflammatory conditions.***
- ***Associated with Secondary amyloidosis.***

Tips to remember: ***It can be remembered through mnemonic ask but written as ASC which means AA is derived from SAA, found in Secondary amyloidosis and seen in chronic conditions and cancers.***

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Beta 2 Microglobulin

A beta2

- ***Abeta 2 Derived from beta 2 microglobulin.***
- ***Associated with CKD.***

Tips to remember: Since there are 2 kidneys so 2 betas for CKD patients.

A beta

- ***Related to beta plaque.***
- ***Associated with Alzheimer's disease***

Tips to remember: Alzheimer's affects the brain, since we have one brain so only beta-amyloid.

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ACal Amyloid

ACAL

- ***Related with calcitonin.***
- ***Associated with Medullary thyroid cancer.***
- ***Tips to remember: Cal (calcitonin) is the Middle (medullary thyroid cancer) man.***

ATTR

- ***Related to transthyretin synthesized by liver.***
- ***Associated with senile systemic amyloidosis.***

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Classification

1. ***Generalized/systemic amyloidosis***

Primary amyloidosis

- ***Amyloid: AL***
- ***Most common type of amyloidosis.***
- ***Seen commonly with plasma cell dyscrasias like multiple melanoma.***
- ***Involves multiple organs.***

Tips to remember: ***P for Primary and Plasma cells, amyloid is also which is India's primary meal and light to eat.***

Secondary amyloidosis

- ***Amyloid: AA***
- ***Associated with TB, Rheumatoid arthritis, Renal cell cancer, Hodgkin lymphoma***

Tips to remember: ***ASC mnemonic can be used.***

Chronic Kidney disease

- ***Amyloid: Abeta2***
- ***Dialysis leads to an increase in serum levels of beta 2 microglobulin.***

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Classification

Senile systemic amyloidosis

- ***Amyloid: ATTR***
- ***Occurs due to the overproduction of Transthyretin.***

2. Localised amyloidosis

Alzheimer's disease

- ***Amyloid: A beta***
- ***Beta plaque is a characteristic feature of Alzheimer's disease.***

Medullary thyroid cancer

- ***Amyloid: Acal***
- ***There is an overproduction of Calcitonin.***

3. Hereditary amyloidosis

Familial Mediterranean fever

- ***Amyloid: AA***

Familial amyloidotic polyneuropathy

- ***Amyloid: ATTR***
- ***Due to mutated Transthyretin.***

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Effects on organs

General effect on organs: there is deposition of a protein in their extracellular space, there will be organomegaly or increase in size seen.

Kidneys

- ***Most commonly affected.***
- ***Amyloid deposits in mesangium cause proteinuria and organomegaly.***

Heart

- ***Amyloidosis is the most common cause of Restrictive cardiomyopathy.***
- ***Amyloid protein deposits in subendocardial tissue leading to arrhythmias***

Liver

Amyloid deposits in the Space of Disse causing hepatomegaly.

Spleen

- ***White pulp involvement: deposit in splenic follicles causing Sago spleen***
- ***Red pulp involvement: deposit in splenic sinuses causing Lardaceous spleen***

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Effects on organs

Skin

Amyloid deposits in the perivascular area lead to pinch purpura.

- ***Joint deposition***
- ***Tongue: deposition leads to macroglossia.***

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Diagnostic methods

(most important)

Congo red stain

- ***Shows apple green birefringence under polarised light.***

Electron microscopy

- ***Shows fibrils***

X ray crystallography

- ***Shows beta pleated structure.***
- ***Imaging (MRI,CT,ECHO) for organ involved.***

Biopsy

- ***For confirmation.***