MAYO CLINIC

Classification of Amyloidosis

Type of amyloidosis	Precursor protein component	Clinical presentation
AL amyloidosis* (previously referred to as primary amyloidosis)	κ or λ immunoglobulin light chain	Primary or localized; see text for syndromes
AA amyloidosis (previously referred to as secondary amyloidosis)	Serum amyloid A protein	Associated with chronic inflammatory conditions; typically acquired, but hereditary in case of familial Mediterranean fever; renal presentation most common
ATTR amyloidosis	Mutated transthyretin	Hereditary; peripheral neuropathy and/or cardiomyopathy
Mutant TTR† (commonly referred to as familial amyloid polyneuropathy)		
Normal TTR† (senile amyloidosis)	Normal transthyretin	Restrictive cardiomyopathy; carpal tunnel syndrome
β2-microglobulin amyloidosis (associated with long-term dialysis)	β2-microglobulin	Carpal tunnel syndrome
Aβ amyloidosis	Aβ protein precursor	Alzheimer syndrome
Other hereditary amyloidosis	Fibrinogen α-chain	Renal presentation
A fibrinogen (also called familial renal amyloidosis)		
Lysozyme	Lysozyme	Renal presentation most common
Apolipoprotein A-I	Apolipoprotein A-I	Renal presentation most common

*AL amyloidosis is the only form of amyloidosis that is secondary to a clonal plasma cell disorder. AL amyloidosis can be associated with multiple myeloma in approximately 10% of patients.

†TTR refers to transthyretin, which is commonly referred to as prealbumin.

From Rajkumar et al Mayo Clinic Proceedings 2006;81(5):693-703