

Amyloidosis (Amyloid light-chain (AL)) (short version)

Recommendations from the society for diagnosis and therapy of haematological and oncological diseases

Publisher

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ICD-10: E85.-

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Compliance rules:

- [Guideline](#)
- [Conflict of interests](#)

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1 Summary

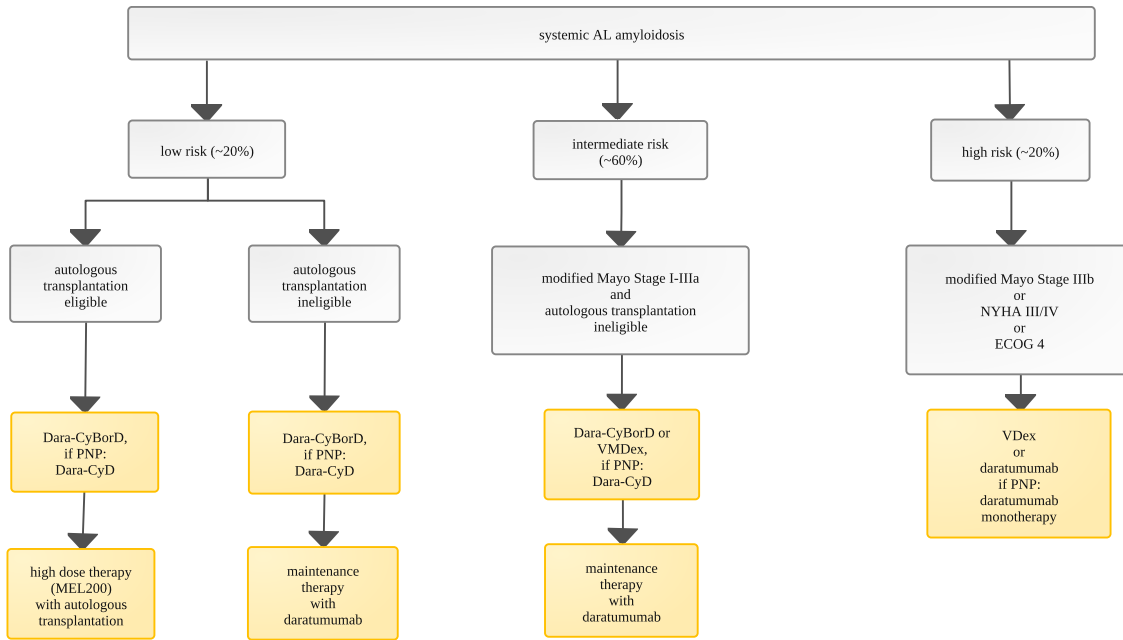
Amyloidoses are rare protein folding diseases in which proteins are deposited as insoluble fibrillar aggregates as a result of a conformational change. This can be systemic (site of production and deposition different) or localized (site of production and deposition identical). Nomenclature is based on the amyloidogenic protein, which has superseded the previous classification into primary and secondary amyloidosis. Systemic amyloidoses are potentially life-threatening complications of monoclonal gammopathies (light chains, AL amyloidoses), age-related diseases (transthyretin, nonmutated; ATTRwt), chronic inflammation (serum amyloid A, AA amyloidoses), or they run in families as part of a monogenetic disease (most commonly transthyretin, TTR amyloidoses). The causal treatment of AL amyloidosis is the reduction of amyloid-forming light chains by immunochemotherapy. Early diagnosis of the disease is essential. At this stage, patients qualify for immuno-chemotherapy and can therefore still be treated effectively, thus avoiding further functional deterioration of the organs.

Patients should be presented to an amyloidosis center before initiating therapy, if possible. A list of centers can be found on the homepage of the German Society for Amyloid Diseases (<http://www.amyloid.de/>). There are also centers in Switzerland and Austria (Swiss Amyloidosis Network of the University Hospital Zurich <https://www.usz.ch/krankheit/amyloidose/>; Interdisciplinary Amyloidosis Center of the Medical University of Vienna).

2 Therapy

The current treatment algorithm is depicted in [Figure 1](#) and [Figure 2](#).

Figure 1: First-line therapy for light chain (AL) amyloidosis



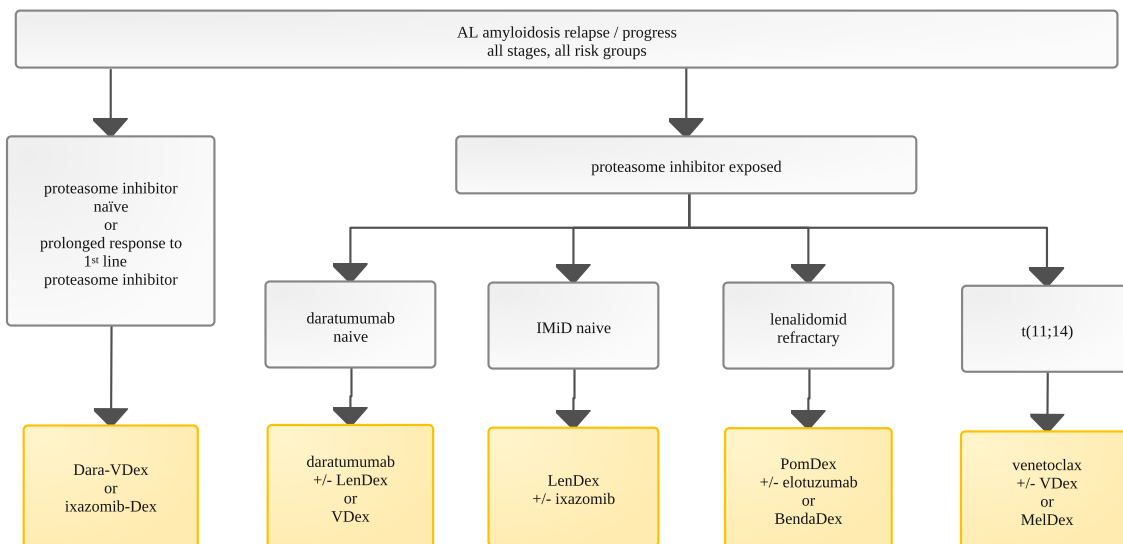
Legend:

— non-curative treatment intent;

For composition and dosages of therapy regimens, see Appendix Therapy Protocols. PNP: polyneuropathy; Dara: daratumumab; Cy: cyclophosphamide; D: Dexamethasone; Boron or V: bortezomib; M or MEL: melphalan.

The risk classification is presented from [chapter 6.1.1.1](#) onwards (link to German version).

Figure 2: Therapeutic options for relapsed/refractory light chain (AL) amyloidosis



Legend:

— non-curative treatment intent;

For composition and dosages of the therapy regimens, see Appendix Therapy Protocols. Dara: daratumumab; Cy: cyclophosphamide; Dex: dexamethasone; V: bortezomib; Beda: bendamustine; Mel: melphalan; Len: Lenalidomide; Pom: Pomalidomide

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16 Disclosures

Conflicts of interest can be found in the [full German version of the guideline](#).