

Amyloidosis Response Criteria

Hematologic Response

Complete Response

Requires **all** of the following:

- Serum and urine negative for monoclonal proteins by immunofixation
- Normal serum free light chain ratio

Very Good Partial Response

- Reduction in the difference between the involved and uninvolved serum free light chain to < 40 mg/L.

Partial Response (PR)

Requires **any** of the following:

- ≥ 50% reduction in serum monoclonal protein levels (if > 0.5 g / dL or > 5 g / L at baseline)
- ≥ 50% reduction in urine light chains (if >100 mg / day at baseline)
- ≥ 50% decrease in clonal serum free light chain levels (if >10 mg / dL or >100 mg / L at baseline)

No Response (NR)/Stable Disease (SD)

Does not meet the criteria for CR, PR, or progressive disease.

Progressive Disease (PD)

Requires **any** of the following:

- If progressing from CR, any detectable monoclonal protein or abnormal free light chain ratio (light chain must double)
- If progressive from PR or SD, ≥ 50% increase in the serum M protein to > 0.5 g/dL, or ≥ 50% increase in urine M protein to > 200mg/day with visible peak present.
- Serum free light chain increase of ≥ 50% to > 10 mg/dL (100 mg/L)

Cardiac Response

Cardiac Response

Requires **any** of the following:

- ≥ 2 mm decrease in mean intraventricular septal wall thickness by echocardiogram
- ≥ 20% increase in left ventricular ejection fraction
- ≥ 2 grade decrease in [New York Heart Association functional class](#) without an increase in diuretic use

and no increase in wall thickness

- Reduction ($\geq 30\%$ and $\geq 300\text{ng/L}$) of NT-proBNP in patients whom the eGFR is ≥ 45 ml/minute/1.73m²

No Response/Stable Disease

Does not meet the criteria for cardiac response or progressive disease

Progressive Disease

Requires **any** of the following:

- ≥ 2 mm increase from baseline in the intraventricular wall thickness by echocardiogram
- $\geq 10\%$ decrease in the left ventricular ejection fraction.
- ≥ 1 grade increase in [New York Heart Association functional class](#)

Hepatic Response

Hepatic Response

Requires **all** of the following:

- ≥ 2 cm decrease in liver span if hepatomegaly present (liver > 15 cm)
- $\geq 50\%$ decrease and/or normalization of serum alkaline phosphatase (ALP) level

No Response/Stable Disease

Does not meet the criteria for hepatic response or progressive disease

Progressive Disease

Requires the following:

- $\geq 50\%$ increase in the serum alkaline phosphatase (ALP) level

Autonomic Neuropathy Response

Autonomic Neuropathy Response

Resolution of symptomatic orthostatic hypotension

No Response/Stable Disease

Does not meet the criteria for autonomic neuropathy response or progressive disease

Progressive Disease

Worsening of symptomatic orthostatic hypotension

Peripheral Neuropathy Response

Peripheral Neuropathy Response

Requires **any** of the following:

- Resolution of abnormal physical findings
- Resolution or improvement of abnormal electromyography (EMG) and/or Nerve Conduction Velocity (NCV) findings

No Response/Stable Disease

Does not meet the criteria for peripheral neuropathy response or progressive disease

Progressive Disease

Requires **any** of the following:

- Worsening of physical findings
- Worsening of EMG and/or NCV findings

Renal Response

Renal Response

- $\geq 50\%$ decrease of at least 0.5 g/day (500mg/24hr) in 24-hour urine protein of > 0.5 g/day (500mg/24hr) pre-treatment **and**
- Creatinine clearance or serum creatinine must not have worsened by $\geq 25\%$ over baseline

If only serum creatinine is obtained, an estimated creatinine clearance can be calculated using the following formula:

$$\text{Estimated Creatinine Clearance} = [(140 - \text{Age (years)}) * \text{Weight (kg)}] / [72 * \text{Serum Creatinine (mg/dL)}]$$

The calculation should be multiplied by 0.85 for women

No Response/Stable Disease

Does not meet the criteria for renal response or progressive disease

Progressive Disease

Requires **any** of the following:

- $\geq 50\%$ increase of at least 1 g/day (1000mg/24hr) for urine protein to $> 1\text{g/day}$ (1000mg/24hr)
- 25% worsening of serum creatinine or creatinine clearance

Manual Updates:

Sections of the Forms Instruction Manual are frequently updated. The most recent updates to the manual can be found below. For additional information, select the manual section and review the updated text.

If you need to reference the historical Manual Change History for this form, please [click here](#) or reference the retired manual section on the [Retired Forms Manuals](#) webpage.

Date	Manual Section	Add/Remove/Modify	Description
5/26/ 21	Amyloidosis Response Criteria	Modify	The Partial Response criteria for Hematologic Response was updated to be consistent with the amyloid response criteria: <ul style="list-style-type: none"> * $\geq 50\%$ reduction in current serum monoclonal protein levels (if $> 0.5\text{ g / dL}$ or $> 5\text{ g / L}$ at baseline) $> 0.5\text{ g/dL}$ * $\geq 50\%$ reduction in urine light chains (if $>100\text{ mg / day}$ at baseline) $\geq 50\%$ reduction in current urine m-protein levels $> 100\text{ mg/day}$ with a visible peak * $\geq 50\%$ decrease in clonal serum free light chain levels (if $>10\text{ mg / dL}$ or $>100\text{ mg / L}$ at baseline) $\geq 50\%$ reduction in current free light chain levels $> 10\text{mg/dL}$
12/ 22/ 20	Amyloidosis Response Criteria	Add	Clarified in the Hematologic Response criteria that free light chain ratios are for the serum .
8/4/ 2020	Amyloidosis Response Criteria	Remove	Removed the following (struck out below) criteria from the Hematologic Complete Response Criteria: <i>Requires all of the following:</i> <ul style="list-style-type: none"> • Serum and urine negative for monoclonal proteins by immunofixation • Normal free light chain ratio • Plasma cells in marrow $< 5\%$
4/6/ 2020	Amyloidosis Response Criteria	Add	Added guidance on Free Light Chain Ratios.
4/19/ 19	Amyloidosis Response Criteria	Modify	Modified the following sections (removed text is struck out below, added text is in red) of the Amyloidosis Response Criteria: Partial Response: <ul style="list-style-type: none"> • $\geq 50\%$ reduction in current serum monoclonal protein levels $> 0.5\text{ g/dL}$ • $\geq 50\%$ reduction in current urine light chain urine m-protein levels

			<p>> 100 mg/day with a visible peak</p> <ul style="list-style-type: none"> • ≥ 50% reduction in current free light chain levels > 10mg/dL <p>Renal Response:</p> <ul style="list-style-type: none"> • ≥ 50% decrease of at least 0.5 g/day (500mg/24hr) in 24-hour urine protein of > 0.5 g/day (500mg/24hr) pre-treatment and • Creatinine clearance or serum creatinine must not have worsened by ≥ 25% over baseline <p><i>If only serum creatinine is obtained, an estimated creatinine clearance can be calculated using the following formula:</i></p> <p>Estimated Creatinine Clearance = [(140 – Age (years)) * Weight (kg)] / [72 * Serum Creatinine (mg/dL)]</p> <p><i>The calculation should be multiplied by 0.85 for women.</i></p>
12/3/18	Amyloidosis Response Criteria	Add	<p>Added further clarification for the Renal Response criteria (indicated in red below):</p> <ul style="list-style-type: none"> • ≥ 50% decrease of at least 0.5 g/day (500mg/24hr) in 24-hour urine protein of > 0.5 g/day (500mg/24hr) pre-treatment and • Creatinine clearance must not have worsened by ≥ 25% over baseline

Last modified: May 26, 2021

New York Heart Association Function Classifications

Class I

Able to perform ordinary activities without symptoms; no limitation of physical activity

Class II

Ordinary physical activity produces symptoms; slight limitation of physical activity

Class III

Less-than-ordinary physical activity produces symptoms; moderate limitation of physical activity

Class IV

Symptoms present even at rest; severe limitation of physical activity

Last modified: Mar 03, 2015