Chief Complaint

“Abdominal pain and swelling x 4-6 weeks”
History of Present Illness:

- 39 year old man with a history of HTN, DMII (dx 10 months prior) was in his usual state of health until...
  - 4 months prior to admission:
    - Noticed fatigue and intermittent nausea and vomiting (initially patient associated symptoms to his recently diagnosed DM, so he ceased metformin use)
    - Began to notice unintentional weight loss
  - 4 weeks prior to admission:
    - Nausea and non-bilious non-bloody vomiting progressed over the prior four months to an unbearable daily occurrence
    - Onset of diffuse abdominal pain and distension for which he presented to an outlying facility
    - Abdominal pain was described as a constant “nagging and aching” not associated with PO intake or BMs
HPI Continued:

- 3 weeks prior to admission:
  - Admitted to OSH x 1 week
  - Found to have hepatomegaly and ascites
  - HCV Antibody Positive
  - U/S guided paracentesis performed (1 liter removed, SAAG 1.0, 200 WBCs, 67% neutrophils, 575 RBCs, culture negative at discharge)
  - Received piperacillin-tazobactam throughout hospital course
  - Underwent liver biopsy, pathology pending at discharge
  - Discharged with spironolactone, furosemide and lisinopril
  - Told to follow up with ILH
HPI Continued:

• 2 days prior to admission:
  • Noticed diffuse pruritis concentrated in bilateral lower extremities and around abdomen, associated with a maculo-papular rash
  • Abdominal pain worsened and was now associated with a 3 day history of midsternal, achy, constant chest pain without radiation or diaphoresis
  • Pain was not associated with exertion but was associated with some mild SOB
Past History

- Past Medical History:
  - Per HPI

- Past Surgical History:
  - None

- Allergies:
  - Tramadol- rash
Past History

- Medications
  - Lisinopril 20mg daily
  - Metformin 500mg BID (None x 4 months)
  - Furosemide 80mg BID
  - Spironolactone 200mg daily

- Family History
  - Father died of lung cancer at 65
  - Mother alive with HTN
Past History

• Social history
  • Current ½ ppd tobacco smoker x 22 years
  • Occasional marijuana use
  • Denies EtOH
  • Monogamous with wife x 20 years
  • Tattoos: All at professional tattoo parlors
    • R arm (1985), L arm (1993), and chest (2008)

• Health Maintenance
  • Not UTD on influenza, pneumococcal, or tetanus immunizations
Review of Systems

- Constitutional: Positive for fatigue and weight loss of 80lbs over past 4 months. Denied fevers or chills.
- Eyes: negative for redness and visual disturbance.
- Ears, nose, mouth, throat, and face: Negative for epistaxis and sore throat.
- Respiratory: Negative for cough, sputum production, hemoptysis, Positive for SOB x10 days.
- Cardiovascular: Positive for chest pain as in HPI.
- Gastrointestinal: Positive for nausea, vomiting, and increased abdominal girth, BRBPR on paper after straining.
- Genitourinary: negative for dysuria and change in frequency.
- Integument: Positive for rash as in HPI. Multiple tattoos.
- Hematologic/lymphatic: negative for bleeding and lymphadenopathy.
- Neurological: negative for weakness, dizziness, gait problems and headaches.
Vital Signs

- BP: 95/79 mmHg
- Pulse: 109/min
- Temp: 98.8 °F
- Resp: 10/min
- SaO2: 92% on RA
- Weight: 123kg
- Height: 6’2”
- BMI: 34
Physical Exam

- General appearance: Ill appearing, appears stated age and cooperative. Resting comfortably
- Head: Temporal wasting otherwise normocephalic, without obvious abnormality, atraumatic
- Eyes: Conjunctivae/corneas clear; minimal scleral icterus; PERRL, EOM's intact.
- Oropharynx: Clear without lesions, mucous membranes minimally dry
- Neck: No adenopathy, no carotid bruit, JVP 7cm, supple, symmetrical, trachea midline and thyroid not enlarged, symmetric, no tenderness/mass/nodules
Physical Exam

- **Lungs**: CTA bilaterally. No Dullness to percussion. No changes in tactile fremitus were appreciated.

- **Heart**: regular rate and rhythm, S1, S2 normal, no murmur, click, rub or S3/S4

- **Abdomen**: Distended; bowel sounds normal; TTP greatest in the RUQ and RLQ; no masses, Liver span 14cm at right midclavicular line by percussion; (+) fluid wave and shifting dullness. No caput medusae or spider angiomas. +Splenomegaly

- **Extremities**: Atraumatic; pitting edema just proximal to bilateral knees and at sacrum

- **Pulses**: 2+ and symmetric

- **Skin**: Skin color, texture, turgor normal. Excoriations bilateral calves and abdomen.
Labs

- WBC: 27.9 (4.5-11)
- HGB: 19 (13.5-17.5 gm/dL)
- HCT: 58 (40-51%)
- PLT: 340
- MCV: 84.2
- RDW: 17.8 (11.5-14.5%)
- NEUTROPHILS: 88
- LYMPHOCYTES: 8
- MONOCYTES: 4
<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Reference Range</th>
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<tbody>
<tr>
<td>NA</td>
<td>138</td>
<td>(3.6-5.2)</td>
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<tr>
<td>K</td>
<td>7.5</td>
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<tr>
<td>CL</td>
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<tr>
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<tr>
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<tr>
<td>GLU</td>
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<td>CALCIUM</td>
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<tr>
<td>Anion Gap</td>
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</tr>
<tr>
<td>Trop</td>
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</tr>
<tr>
<td>BNP</td>
<td>89</td>
<td></td>
</tr>
<tr>
<td>PROTEIN</td>
<td>5.4</td>
<td>(6-8)</td>
</tr>
<tr>
<td>ALBUMIN</td>
<td>1.8</td>
<td>(3.4-5)</td>
</tr>
<tr>
<td>TOTAL BILI</td>
<td>5.1</td>
<td>(&lt;1.3)</td>
</tr>
<tr>
<td>AST</td>
<td>80</td>
<td>(&lt;45)</td>
</tr>
<tr>
<td>ALK PHOS</td>
<td>1674</td>
<td>(20-120)</td>
</tr>
<tr>
<td>ALT</td>
<td>51</td>
<td>(&lt;46)</td>
</tr>
<tr>
<td>Lactic Acid</td>
<td>6.1</td>
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<td>PT</td>
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<td>(9.2-12.5 sec)</td>
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<td>FT4</td>
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</table>
Labs

- Urinalysis
  - Protein  75
  - Bili  +

- Urine, Microscopic
  - RBC  3-5 (0-2/hpf)
  - WBC  3-5
  - SE  2-20
  - Bacteria  Neg
  - Cast  0-2
Abdominal X-ray at Presentation
Abdominal Ultrasound at Presentation

- Hepatosplenomegaly (liver 25 cm, spleen 15 cm)
- Edematous liver
- Mildly enlarged kidneys
- Moderate volume abdominal ascites
- Mild left hydronephrosis
Hospital Course - Day 1

• Day 1:
  • Admitted to ICU and started on treatment for hyperkalemia; ceftriaxone for SBP coverage; and volume resuscitation
  • Blood and Urine Cultures drawn
  • Nephrology Consult:
    • Upon further record review, patient with normal Creatinine at outside facility
    • Spun urine showed numerous granular and muddy brown cast and WBCs
    • SPEP, UPEP ordered
    • DDX: ATN vs Inflammatory vs Amyloid
Hospital Course- Day 2

• Day 2:
  • Labs:
    • Potassium- 5.5
    • Lactic Acid- 1.8
    • WBC- 24
    • INR- 1.7
    • Hepatitis C Antibody- Positive
  • Stepped down to Medicine service
  • Failed attempt at bedside diagnostic paracentesis => IR consulted
Hospital Course - Day 3

- Day 3:
  - Pathology Report returned from OSH inconclusive, reviewed by Tulane Pathologist:
    - *Amyloidosis*, with extensive amyloid deposition in space of Disse and in portal vascular branches. The interpretation of amyloidosis is based on the pattern of deposits (perisinusoidal and vascular deposition) and the Congo Red staining showing characteristic apple-green birefringence
    - Portal-to-Portal bridging fibrosis
    - Mild Iron deposition in hepatocytes (2+ positivity on a scale of 0-4)
Google Image - Lung with Apple-Green Birefringence
Hospital Course - Day 3

- Day 3:
  - Confirmatory test for Hepatitis C: Negative
    - HCV antibody verification non-reactive
    - Hepatitis C RNA-PCR: HCV RNA not detected
  - Blood Cultures Positive for *Streptococcus pneumoniae*
  - Diagnostic paracentesis: Performed by IR
    - WBC 15,218 (Segs 94%, Lymphs 4%, Monos 4%), RBC 1122
    - Albumin <1, Bili 0.5, Protein <3, glucose 101, Amylase 11
Hospital Course - Day 3

• Day 3 Continued:

• GI Consult:
  • RUQ with doppler to evaluate hepatic venous flow
  • Daily INR
  • Continued Ceftriaxone
  • Alpha-1 Antitrypsin 236 (90-200)
  • Ceruloplasmin 39 (15-30)

• Heme Onc Consult:
  • Recommended following up SPEP/UPEP, serum free light chains, cardiac MRI, TTR mutation analysis; plans made for bone marrow biopsy
Hospital Course- Day 5

● Day 5:
  ● Cardiology Consult:
    • Most likely subclinical myocardial amyloid deposits based on EKG findings of small QRS voltage with pseudo-infarction (anterior and inferior) and RVH
    • TT Echocardiogram: no signs of restrictive cardiomyopathy
    • Could not undergo Cardiac MRI secondary to renal failure
    • Myocardial Biopsy not warranted
Hospital Course- Days 5-9

- Days 5-9:
  - Continued treatment for *S. pneumoniae* bacteremia
  - Increased urine output, Creatinine trended down to 3.19 and potassium stabilized within normal limits
  - WBC ranged between 16-29K
  - Further cultures remained NGTD
  - Concern for superimposed LE cellulitis => treated with vancomycin and ciprofloxacin
Laboratory Results

- **UPEP:**
  - All major protein fractions identified in a pattern suggestive of glomerular protein loss. No abnormal protein bands identified.

- **SPEP:**
  - Pattern consistent with non-specific hypoalbuminemia

- **Free Kappa Light Chain:** 852.5 (3.3-19.4 mg/L)

- **Free Lambda Light Chain:** 25.6

- **Kappa/Lambda Ratio:** 33.3 (0.26-1.65 mg/L)
Bone Marrow Core Biopsy
Bone Marrow- CD 138 + Plasma Cell Infiltrate
Bone Marrow- Congo Red Stain
Bone Marrow Aspirate- Immature Plasma Cells
Bone Marrow Core - Kappa
Bone Marrow Core - Lambda
Hospital Course – Discharge

- Per patient request, discharged after 14 days

Follow up:

- 4 days after discharge with Hematology/Oncology for initiation of chemotherapy with CyBorD (Cyclophosphamide, Bortezomib, Dexamethasone) dose-reduced for liver and renal dysfunction with potential stem cell transplant after chemotherapy
- 1 week after discharge with Nephrology
- 1-2 after discharge with Gastroenterology
Hospital Course – Readmit

- Seen in Oncology clinic 4 days later, sent to ED due to hypotension (BP 99/54 in clinic), generalized weakness, abdominal and lower extremity swelling, abdominal tenderness and dyspnea on exertion

- In ED MICU consulted for BP 70s/40s and abnormal lab work:
  - WBC- 16 with a normal differential, H&H and platelet count
  - BUN/Creatinine- 77/6.16 (at discharge 54/4.13)
  - Bicarb- 19
  - K- 5.7
  - Bilirubin- 7.7, AST- 127, Alk Phos- 1470, ALT- 148 (all elevated from discharge)
  - Urinalysis- Amber, 25 blood, 3-5 RBCs, 500 protein, 500 leukocytes, >100 WBCs, 1-2 squam, many bacteria, WBC clumps, hyaline casts, course granular casts

- Admitted to MICU with severe sepsis secondary to UTI, treatment with IV vancomycin, piperacillin-tazobactom and ciprofloxacin as well as IVFs
Abdominal CT Without IV Contrast - Readmit
Abdominal CT Without IV Contrast - Readmit
Amyloidosis- Radiology

- Nonspecific imaging
- Hepatomegaly
- Focal low attenuation in liver and spleen
  - Amyloid pseudotumor
- Delayed enhancement
Acute Hepatitis/Hepatotoxicity - Radiology

- Normal liver is hypointense to the spleen on T2 (and hyperintense to the spleen on T1)
- This liver is diffusely hyperintense on T2
Acute Hepatitis/Hepatotoxicity

• US - “Starry sky”
• Appearance of the liver on grayscale ultrasound
  • Swelling of hepatocytes causes decreased echogenicity of the liver parenchyma with better visualization of the fibrous walls of the portal veins
  • CT – very low attenuation on non-contrast; heterogeneous enhancement
Hospital Course – Readmit

- Renal Consult:
  - Due to oliguria and hyperkalemia => CRRT initiated
- Gastroenterology Consult:
  - Patient developed AMS, initiated rifaximin and lactulose
  - Continue broad spectrum antibiotics
- Hematology-Oncology Consult:
  - Initially not a candidate for chemotherapy due to active infection
Hospital Course – Readmit

- 4 Days after readmit patient remained on CRRT
- Hematology-Oncology initiated therapy with Dexamethasone
  - No Cyclophosphamide due to renal failure
  - No Bortezumib as dialyzable and would not get appropriate levels with CRRT due to dose reduction in setting of liver dysfunction
- Patient’s clinical course remained complicated throughout his 22 day admission
- Ultimately, patient intubated due to respiratory failure
- After multiple family discussions patient made DNR and died shortly thereafter
Final Diagnosis

- AL Amyloidosis and Multiple Myeloma
THANK YOU
Amyloidosis

- Generic term that refers to the extracellular tissue deposition of fibrils composed of low molecular weight subunits of a variety of normal serum proteins
  - In antiparallel β-pleated sheet configuration
    - Identified on biopsy specimens both by:
      - Characteristic appearance on electron microscopy
      - Ability to bind:
        - Congo red
          - Leading to green birefringence under polarized light
        - Thioflavine T
          - Producing an intense yellow-green fluorescence.
This Congo red stain reveals amorphous orange-red deposits of amyloid, which is an abnormal accumulation of breakdown products of proteinaceous material that can collect within cells and tissues. 

[http://library.med.utah.edu/WebPath/CINJHTML/CINJ037.html](http://library.med.utah.edu/WebPath/CINJHTML/CINJ037.html)
amyloid in the kidney, thioflavin T (stains amyloid yellow and everything else black)

http://www.pathguy.com/lectures/imm-iii.htm
Amyloidosis

- >20 distinct low molecular weight proteins recognized.
  - Two most common causes of systemic amyloid deposition are:
    - Immunoglobulin light chain (AL) amyloidosis (previously primary amyloidosis)
      - Fibrils composed of fragments of monoclonal light chains.
      - May have amyloidosis alone or in association with other plasma cell dyscrasias
        - Multiple myeloma, Waldenstrom macroglobulinemia
    - All forms of systemic amyloidosis derived from monoclonal light chains are considered AL amyloidosis.
  - AA amyloidosis
    - Composed of fragments of the acute phase reactant serum amyloid A.
    - AA amyloidosis is typically reactive (secondary) to chronic inflammation
Epidemiology of AL disease

- In the United States, the incidence appears to be stable
  - Approximately 6 to 10 cases per million person-years
  - Exact incidence is unknown.

- Disease of older adults
  - Age-specific incidence increase in each decade of life after 40
  - Median age at diagnosis is 64 years
  - Less than 5 percent of patients are under the age of 40
  - Male predominance
    - Account for 65 to 70 percent of patients.

- Occurs in all races and all geographic locations
  - Little data regarding whether the incidence varies by ethnicity or geography.
Association with other disorders

• Can present with
  • Plasma cell dyscrasias including:
    • Monoclonal gammopathy of undetermined significance (MGUS)
    • Multiple myeloma
    • Waldenström macroglobulinemia
  • Malignant disorders of:
    • Plasma cells
    • Lymphoplasmacytic cells
Above Patient association

- Myeloma typically diagnosed before or around the time of the amyloid diagnosis.
  - Less commonly, myeloma develops more than six months after the diagnosis of amyloid (delayed progression)
- These findings are suggestive of the combination of myeloma and amyloidosis:
  - 30 percent or more plasma cells
  - Hypercalcemia
  - Bone pain
  - Or lytic bone lesions
Clinical Presentation

• Depends on the number and nature of the organs affected
• Non-specific common systemic symptoms include:
  • Fatigue
  • Unintentional weight loss
• Other common clinical presentations
  • Nephrotic syndrome
  • Restrictive cardiomyopathy
  • Peripheral neuropathy
  • Hepatomegaly with elevated LFT’s
  • Macroglossia and involvement of other muscles
  • Purpura and other skin manifestations
  • Bleeding diathesis
Evaluation

- Suspect in a patient presenting with any one of the following:
  - Non-diabetic nephrotic range proteinuria
  - Restrictive cardiomyopathy or otherwise unexplained CHF
    - Increased NT-proBNP in the absence of primary heart disease
  - Unexplained edema
  - Hepatosplenomegaly
  - Carpal tunnel syndrome
  - Unexplained facial or neck purpura
  - Or macroglossia.
Evaluation and Diagnosis

• Initial evaluation of a pt suspected of AL amyloidosis should consist of:
  • Serum and urine immunofixation
  • Serum free light chain ratio analysis
  • Abdominal fat pad aspirate and bone marrow biopsy.
    • In the setting of negative fat pad aspirate and bone marrow biopsy and high suspicion
    • Affected organ (eg, kidney, liver) should be biopsied.

• Diagnosis does requires a biopsy.
  • Preferred sites:
    • Abdominal fat pad aspirate
    • Bone marrow biopsy
    • Either or both are positive in 90 percent of patients with AL amyloidosis.
Diagnosis cont.

• Require the presence of all of the following:
  - Presence of an amyloid-related systemic syndrome
    - Renal, liver, heart, gastrointestinal tract or peripheral nerve involvement
  - Positive amyloid staining by Congo red in any tissue
    - Fat aspirate, bone marrow or organ biopsy
  - Evidence that the amyloid is light chain-related established by direct examination of the amyloid
    - Immunohistochemical staining, Immunofluorescence microscopy, Amino acid sequence analysis or Laser microdissection with mass spectrometry
  - Evidence of a monoclonal plasma cell proliferative disorder
    - Presence of a serum or urine M protein, abnormal serum free light chain ratio, or clonal plasma cells in the bone marrow).
Treatment of AL Amyloidosis

- To best treat patients the initial evaluation must confirm:
  - Confirm diagnosis
  - Establish the extent and sites of disease, and
  - Evaluate for comorbidities likely to impact treatment options

- Initial management varies if patients are eligible to pursue high dose melphalan followed by autologous hematopoietic cell transplantation (HCT).

- Patients not considered transplant candidates with:
  - Poor performance status
  - Major comorbidities
  - Involvement of three or more organs, and
  - Advanced cardiac amyloidosis
HCT eligible patients

- Treat with high dose melphalan followed by hematopoietic stem cell rescue (Grade 2C).
- With better patient selection and using a risk-adapted approach
  - Results with HCT may be superior to those obtained following chemotherapy alone.

Approach to therapy in patients with AL amyloidosis eligible for hematopoietic cell transplantation (HCT)

- Newly diagnosed AL amyloidosis - transplant eligible
  - HCT with melphalan
    - <90 percent reduction in dFLC at day +100
    - ≥90 percent reduction in dFLC at day +100
      - High risk
        - <50 percent reduction in dFLC
          - Bortezomib-based therapy
        - ≥50 percent reduction in dFLC
          - Observe
      - Standard risk
        - Observe
  - Observe

dFLC: difference between involved and uninvolved serum free light chain levels.
* High risk: Mayo stage III (cTnT >0.035 mcg/L and NT-proBNP >332 ng/L).
* Start alternate therapy if organ progression at any time.
Reproduced with permission from: mSMART (msmart.org).
Not eligible for HCT

- Treat with melphalan/dexamethasone (Grade 1C).
- High risk for complications due to Mayo Stage III cardiac involvement (cardiac troponin >0.035 and NT-proBNP >332) are switched to:
  - Bortezomib-based therapy if:
    - Failure to attain at least a partial response by six weeks or
    - Very good partial response by three months.
- Patients who are Stage I or II are switched to:
  - Bortezomib-based regimens if:
    - Failure to attain at least a partial response by three months or
    - Very good partial response by six months.
Approach to therapy in patients with AL amyloidosis not eligible for hematopoietic cell transplantation (HCT)

Newly diagnosed AL amyloidosis - transplant ineligible

Melphalan plus dexamethasone

High risk*

≥50 percent reduction in dFLC at six weeks* and ≥90 percent reduction in dFLC at three months*

Yes → Observe* No → Bortezomib-based therapy

≥50 percent reduction in dFLC at six months*

No → Observe* Bortezomib-based therapy

Standard risk

≥50 percent reduction in dFLC at three months* and ≥90 percent reduction in dFLC at six months*

Yes → Observe* No → Bortezomib-based therapy

dFLC: difference between involved and uninvolved serum free light chain levels.
* High risk: Mayo stage III (cTnT >0.035 mcg/L and NT-proBNP >332 ng/L).
- Start alternate therapy if organ progression at any time.
Reproduced with permission from: mSMART (msmart.org).
Relapsed disease

- Bortezomib-based or immunomodulatory-based regimens is a reasonable approach.
- No good data to determine which of these regimens will be of most benefit
- Choice will be dictated by:
  - Patient and physician preferences
  - Expected toxicity
  - Drug availability
  - Insurance coverage.
Response Assessment

- Response to tx should be documented 3 months following the completion of planned therapy or sooner if the outcome is unfavorable by:
  - History and physical examination
  - Laboratory studies
    - Serum free light chain assay
    - NT-proBNP
    - 24-hour urinary protein and creatinine clearance
    - Electrophoresis of the serum and urine
    - Immunelectrophoresis of the serum and urine
    - Alkaline phosphatase
  - Other diagnostic studies
    - Echocardiogram,
    - Radiographic measurement of the liver
    - EMG
    - Nerve conduction studies
## Prognostic staging systems for amyloidosis

<table>
<thead>
<tr>
<th>Prognostic model</th>
<th>Risk groups</th>
<th>Survival in patients not undergoing stem cell transplantation</th>
<th>Survival in patients undergoing stem cell transplantation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Median in months</td>
<td>Five-year survival rate (percent)*</td>
<td>Median in months</td>
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<tr>
<td><strong>Mayo stage 1</strong></td>
<td>Carcic troponin &lt;0.035 mcg/L and NT-proBNP &lt; 332 ng/L</td>
<td>26</td>
<td>28</td>
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<tr>
<td><strong>Stage II</strong></td>
<td>Any one factor high</td>
<td>11</td>
<td>12</td>
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<tr>
<td><strong>Stage III</strong></td>
<td>Carcic troponin ≥0.035 mcg/L and NT-proBNP ≥ 332 ng/L</td>
<td>4</td>
<td>6</td>
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<tr>
<td><strong>Stage IV</strong></td>
<td>NT-proBNP &lt;1800 ng/L, carcic troponin T &lt;0.025 mcg/L, and difference between involved and uninvolved serum free light chains &lt;18 mcg/dL</td>
<td>55</td>
<td>50</td>
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<tr>
<td><strong>Stage V</strong></td>
<td>Any one factor high</td>
<td>19</td>
<td>35</td>
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<tr>
<td><strong>Stage VI</strong></td>
<td>Any two factors high</td>
<td>12</td>
<td>20</td>
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* Estimated from published survival curve if not reported.
* Survival rate at 40 months.

**References:**
<table>
<thead>
<tr>
<th>Prognostic model</th>
<th>Risk groups</th>
<th>One-year mortality rate (percent)</th>
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<tr>
<td>Early mortality score[1]</td>
<td>Low risk: Cardiac troponin ≤0.01 microg/L, NT-proBNP ≤4200 ng/L, and serum uric acid ≤8.0 mg/dL</td>
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<td>Low-intermediate risk</td>
<td>Any one factor high</td>
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<tr>
<td>High-intermediate risk</td>
<td>Any two factors high</td>
<td>61</td>
</tr>
<tr>
<td>High risk</td>
<td>Cardiac troponin T level &gt;0.01 microg/L, NT-proBNP &gt;4200 ng/L, and serum uric acid &gt;8.0 mg/dL</td>
<td>80</td>
</tr>
</tbody>
</table>

References:

Rajkumar, VS; Clinical presentation, laboratory manifestations, and diagnosis of immunoglobulin light chain (AL) amyloidosis (primary amyloidosis); UpToDate; 5/2013
Rajkumar, VS; Prognosis and treatment of immunoglobulin light chain (AL) amyloidosis and light and heavy chain deposition diseases; UpToDate; 12/2013