Systemic Diseases and the Kidney

FIGURE 3-7
Symptoms of primary systemic amyloidosis in patients during an 11-year study at the Mayo Clinic. Weakness or fatigue and weight loss were the most frequent initial symptoms seen within 30 days of diagnosis. Weight loss occurred in more than half of patients. The median weight loss was 23 lb; five patients lost more than 100 lb each. Purpura, particularly in the periorbital and facial areas, was noted in about one sixth of patients. Gross bleeding was reported initially in only 3%. Skeletal pain was a major symptom in only 5% and usually was related to lytic lesions or fractures associated with multiple myeloma. Dyspnea, pedal edema, paresthesias, light-headedness, and syncope were noted. (From Kyle and Gertz [5]; with permission.)

FIGURE 3-8
Macroglossia in a man with primary systemic amyloidosis. Macroglossia occurs initially in about 10% of patients. Note the imprint of the teeth on the dorsum of the tongue. This patient was unable to close his mouth and complained of drooling. Macroglossia may cause obstruction of the airway, sometimes necessitating a tracheostomy. (From Kyle [4]; with permission.)

FIGURE 3-9
Nodules causing occlusion of the auditory canal in a patient with primary systemic amyloidosis. The external auditory canal may be occluded completely by nodules of amyloid. This condition frequently produces deafness, which may be the initial symptom. (From Gertz and Kyle [6]; with permission.)

FIGURE 3-10
Shoulder pad sign in a woman with primary systemic amyloidosis. Infiltration of the periarticular tissues with amyloid may produce this sign. The shoulder pad sign causes pain and limitation of motion and is very difficult to treat. (From Kyle [4]; with permission.)
Hypertrophic form of primary systemic amyloidosis in a 39-year-old man with prominent and firm muscles. Despite the muscular appearance, results of a biopsy revealed displacement of muscle fibers with amyloid. Patients often exhibit stiffness or limitation of movement. (From Kyle and Greipp [7]; with permission.)

![Figure 3-11](hypertrophic_form_of_amyloidosis.png)

Signs of primary systemic amyloidosis in patients during an 11-year study at the Mayo Clinic. The liver was palpable in about one fourth of patients seen within 30 days of diagnosis. Hepatomegaly is due to infiltration of amyloid or congestion from heart failure. The spleen is palpable in only 5% of patients and rarely extends more than 5 cm below the left costal margin. Lymphadenopathy occurs infrequently. (Adapted from Kyle and Gertz [5]; with permission.)

![Figure 3-12](signs_of_amyloidosis.png)

### HEMOGLOBIN AND PLATELET VALUES WITHIN 30 DAYS OF DIAGNOSIS OF PRIMARY SYSTEMIC AMYLOIDOSIS, MAYO CLINIC, 1981–1992

<table>
<thead>
<tr>
<th>Factor</th>
<th>Median</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin, g/dL (&lt;10 g/dL in 11%)</td>
<td>12.9</td>
<td>6.6–18.6</td>
</tr>
<tr>
<td>Platelets, $\times 10^9$/L (&gt;500 $\times 10^9$/L in 9%)</td>
<td>288</td>
<td>4–953</td>
</tr>
</tbody>
</table>

![Figure 3-13](hemoglobin_platelet_values.png)

Signs creatinine (mg/dL) in patients at diagnosis of primary systemic amyloidosis. Renal insufficiency was present in almost half of patients. Proteinuria was present in about 75% of patients. (Adapted from Kyle and Greipp [7].)
Systemic Diseases and the Kidney

3.6

FIGURE 3-15
Results of serum protein electrophoresis in patients at diagnosis of primary systemic amyloidosis. The serum protein electrophoretic pattern showed hypogammaglobulinemia in 20% of patients. Only half of patients had a localized band or spike in the β or γ areas of the electrophoretic pattern. The median size of the M spike was 1.4 g/dL. In the remaining patients the pattern was normal.

FIGURE 3-16
Serum monoclonal (M-) protein in patients at diagnosis of primary systemic amyloidosis in an 11-year study at the Mayo Clinic. Immunoelectrophoresis or immunofixation of the serum showed an M-protein in 72% of patients. IgG was most common, followed by IgA. Twenty-four percent of patients had monoclonal immunoglobulin light chains in the serum (Bence Jones proteinemia). (Adapted from Kyle and Gertz [5]; with permission.)

FIGURE 3-17
Urine total protein values in patients at diagnosis of primary systemic amyloidosis in an 11-year study at the Mayo Clinic. More than one third of patients exhibited 24-hour urine total protein values of 3.0 g/d or more. Over half of patients had a urine protein value of more than 1 g/d. The electrophoretic pattern showed mainly albumin. (Adapted from Kyle and Gertz [5]; with permission.)

FIGURE 3-18
Urine monoclonal (M-) protein in patients at diagnosis of primary systemic amyloidosis in an 11-year study at the Mayo Clinic. Almost three fourths of patients had monoclonal light chains in their urine on immunoelectrophoresis or immunofixation. In contrast to the type of protein found in multiple myeloma, λ is twice as common as is κ. The 24-hour total amount of monoclonal (M-) protein in the urine was less than 0.5 g/d in more than half of patients. (From Kyle and Gertz [5]; with permission.)

FIGURE 3-19
Serum (S) and urine (U) proteins in patients with primary systemic amyloidosis in an 11-year study at the Mayo Clinic. Immunoelectrophoresis or immunofixation of serum and appropriate concentrations in urine showed a monoclonal protein in nearly 90% of patients. In the absence of monoclonal protein, one must search for a monoclonal population of plasma cells in the bone marrow or perform immunohistochemical staining to identify the type of amyloid. (From Kyle and Gertz [5]; with permission.)

FIGURE 3-20
Enlarged kidney in primary systemic amyloidosis. Involvement of the kidneys is the most common presenting feature. The kidney is frequently normal in size, but in some instances small kidneys have been found.