BENCE JONES PROTEIN IN PRIMARY SYSTEMIC AMYLOIDOSIS

To the Editor:

The recent population-based study by Kyle et al., which was the first to provide precise epidemiologic information about primary systemic amyloidosis, reported the demonstration of a monoclonal (M)-protein in the serum or urine from 16 of 21 patients (76%) with amyloid of unequivocal Ig origin (AL).

However, the number of patients in whom free monoclonal Ig light chain (i.e., Bence Jones [BJ] protein) represented the sole detectable M-protein was apparently lower than I would have expected to find in this disease. The frequency with which BJ proteinuria occurred in association with an intact M-protein in the serum is somewhat difficult to evaluate from the separate presentation of the results of the serum and urine investigations. All but one patient had the serum protein pattern analyzed by at least electrophoresis, with an M-protein band being recognizable in 18 (90%), whereas in only 2 (12%) of the 17 patients who had their serum further investigated by immunoelectrophoresis or immunofixation did the M-protein consist of free \( \lambda \) light chain. The search for BJ protein by immunoelectrophoresis or immunofixation was negative in the urine from one-third of the 15 patients and 3 others had a normal urine electrophoretic pattern. Thus, negative results were obtained in 44% of the 18 patients who had their urine studied by at least electrophoresis. These figures most likely reflect the necessary inclusion in such a retrospective study of patients who had their serum and urine examined by using less sensitive techniques than those available at present. The same reason might also account in part for the 11% to 14% frequency of free light chains detected in the serum. The ability to confidently detect small amounts of urine BJ protein would be of even more interest for the clinician in primary systemic amyloidosis, as the discovery of such an M-protein abnormality does often suggest appropriate investigation and diagnosis.

The recent population-based study by Kyle et al., which was the first to provide precise epidemiologic information about primary systemic amyloidosis, reported the demonstration of a monoclonal (M)-protein in the serum or urine from 16 of 21 patients (76%) with amyloid of unequivocal Ig origin (AL).

Table 1. Serum and Urine M-Proteins in 18 Patients With Primary Systemic Amyloidosis

<table>
<thead>
<tr>
<th>M-Protein</th>
<th>No. of Patients (%)</th>
<th>Serum BJ Protein No.</th>
<th>Urinary BJ Protein Excretion (g/dL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intact M-protein</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IgG ( \kappa )</td>
<td>1 (6)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>IgG ( \lambda )</td>
<td>4 (22)</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>IgA ( \lambda )</td>
<td>2 (11)</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>7 (39)*</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>BJ protein only</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>( \kappa )</td>
<td>4 (22)</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>( \lambda )</td>
<td>7 (39)</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>11 (61)</td>
<td>11</td>
<td>2</td>
</tr>
<tr>
<td>All M-proteins</td>
<td>18 (100)</td>
<td>16*</td>
<td>5</td>
</tr>
</tbody>
</table>

* Serum M-protein concentration (median, 1.4 g/dL; range, 0.5 to 3.5 g/dL).
† Serum BJ protein concentration (median, 0.37 g/dL; range, 0.1 to 0.9 g/dL).
‡ Urinary BJ protein excretion (median, 0.92 g/dL; range, 0.17 to 16.6 g/dL).

REFERENCES

CORRESPONDENCE

RESPONSE

Dr. Pascali has correctly emphasized the importance of finding a monoclonal protein in the serum and urine of patients with primary amyloidosis (AL). As he pointed out, our study began in 1950, when immunoelectrophoresis and immunofixation were not available.

We found that 85% of the 710 AL patients we examined at the Mayo Clinic from 1982 through 1991 had a monoclonal protein in the serum or urine. When we examined the bone marrow for a monoclonal excess of plasma cells in patients without a monoclonal protein in the serum or urine, we found that 98% of patients with AL will have either a monoclonal protein in the serum or urine or a monoclonal excess of bone marrow plasma cells. Immunohistochemical staining of the amyloid is necessary in the remaining patients to differentiate AL from hereditary or senile systemic amyloidosis.

ROBERT A. KYLE
Hematology and Internal Medicine
Mayo Clinic
Rochester, MN

REFERENCE
Bence Jones protein in primary systemic amyloidosis [letter; comment]

E Pascali