Management for Interstitial Lung Disease*
State of the Art
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A survey of 109 pulmonary physicians representing both private practice and academic medicine was conducted to learn their practice of management and treatment of interstitial lung disease. Of the 109, 25 (23 percent) responded. Twenty-two of 23 routinely obtain tissue diagnosis. The routine method was transbronchial biopsy by 15 physicians vs open lung biopsy by seven. Few (n = 5) use BAL cell analysis. The majority of clinicians used two to four criteria to initiate therapy. Physiology laboratory data were the predominant criteria used by the majority of physicians and also were most commonly used to monitor a patient’s course over time. Prednisone was the drug of choice for therapy. Immunosuppressive therapy was not used until failure of steroid therapy was observed. There was little consistency in the diagnostic procedures applied, criteria for the decision to treat, the drugs used to treat, or methods for monitoring.

Diffuse interstitial pneumonitis is a relatively common problem encountered by the pulmonary subspecialist. Substantial investigative effort has been devoted to this entity, and has generated a significant body of literature. However, many fundamental aspects of this disorder remain enigmatic, eg, pathogenesis and natural history. Further, there is a limited literature18 dealing with those aspects of the disorder that the practicing pulmonary physician must address: Which patients should be treated? When? With what? Considering the dearth of information available, we decided to develop a multiphysician study that would allow evaluation of current treatment strategies in this disorder. To establish a suitable protocol, we attempted to collect information about the current approach to such patients. We assumed that there was some consensus regarding the initial evaluation technique employed, the criteria for initiating therapy, the type of therapy employed, and the methods for evaluating response. The first physician population informally surveyed was our own faculty colleagues. This survey quickly disclosed that no common “algorithm” existed. The specific approaches used appeared to reflect the particular investigative interests of the individual and his acceptance or rejection of “the literature.” Many had strong views but agreed that they were difficult to validate because definitive studies were not available.

This informal survey led us to two conclusions: (1) if any management trials were to be initiated, generation of an adequate patient base would require participation by many pulmonary physicians, in and out of full-time academia; (2) a more formal survey of such a population was required to assess current practices with respect to this disorder. This report deals with results of that survey.

MATERIAL AND METHODS
The population selected for survey consisted of former fellows of the UCSD Pulmonary Training Program, a program in existence since 1968. That population was selected because it shared some “common heritage” of training.

A questionnaire was developed to determine the respondents’ interest in participating in a future study, the number of such patients seen per year, and current management practices. With respect to management, the questionnaire specifically requested information regarding: (1) what tests were performed during the initial diagnostic work-up (biopsy material, lavage, cell analysis, blood and/or humoral markers, radiographic and nuclear medicine evaluations); (2) criteria for initiating drug therapy; (3) the drugs used for treatment; (4) what laboratory tests were used to follow patients over time (monitoring course).

Space for comment was provided. Nonrespondents to the first questionnaire were recontacted by a second request six months later. One hundred nine former trainees were solicited. All are certified by the Pulmonary Subspecialty Board.

RESULTS
Twenty-five (23 percent) of those contacted ultimately responded by fully completing the questionnaire. Many others provided a partial response that was not adequate for analysis. However, all respondents indicated a potential interest in participating in a contemplated investigation of this type.

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Table 1—Histologic/Cellular Analysis to Establish the Diagnosis

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Routine Use</th>
<th>Do Not Use</th>
<th>Limited Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Open lung Bx</td>
<td>7</td>
<td>5</td>
<td>11</td>
</tr>
<tr>
<td>Transbronchial Bx</td>
<td>15</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Lavage for BAL cell analysis</td>
<td>5</td>
<td>18</td>
<td>0</td>
</tr>
</tbody>
</table>

Initial Diagnostic Evaluation

With respect to the data base assembled in the initial evaluation of the patient, all respondents included history, physical examination, chest x-ray film, and pulmonary spirometric tests, including Dsb. Few performed arterial blood gas studies at rest or during exercise. Serologic blood tests were rarely obtained. A majority performed transbronchial lung biopsy (TBB): 15 routinely and three on some occasions (Table 1). Open lung biopsy was routinely employed by seven, and on a limited basis by 11. Some responded that open lung biopsy was not performed, even if the TBB was negative unless an infectious etiology was entertained. Others recommended lung biopsy in the clinical setting of a rapidly deteriorating patient. The age of the patient also was a factor: the younger the patient, the more aggressive the work-up, which would include open lung biopsy. Those who did not attempt tissue diagnosis relied on history, symptoms, and the chest roentgenogram.

Only five of the 23 responders did BAL cell analysis. All five are in academic institutions. Of the 18 who do not use BAL cell analysis, six expressed an interest but have problems with cytopathology interpretation at their institution. The remaining 12 do not do BAL analysis because of personal preference.

Criteria for Initiating Drug Therapy

With respect to those criteria used to initiate therapy, there was no unanimity. The number of tests used to decide whom to treat varied from one abnormal test to as many as six. The majority of physicians (n = 14) used two to four criteria for initiating treatment. However, the tests used were individualized to the specific patient and did not follow any fixed pattern. The physiology laboratory findings and the patient's physical findings on examination were the two predominant criteria used by the majority of physicians to initiate therapy (Table 2). Some physicians remarked that they do not initiate drug therapy at the time the diagnosis is made if symptoms are mild, but wait until there is further deterioration in lung function before instituting treatment.

Monitoring Course

Physiologic data were most commonly used to follow patients over time (Table 3). The gallium lung scan, chest x-ray films, and subjective symptoms were relied on fairly equally. Bronchoalveolar lavage was not used to follow a patient's course over time by any respondent.

Drug Therapy

The drug of choice by nearly all the responders was prednisone (n = 18). One used cyclophosphamide as the first-line drug in patients over age 50. One other physician indicated he may use both cyclophosphamide and prednisone in combination as the initial drug therapy regimen.

If the patient deteriorated in the face of prednisone therapy, the most common response was to add a second drug (n = 13), either cyclophosphamide or azathioprine. Two would increase the prednisone dose in conjunction with adding a nonsteroidal, immunosuppressive drug. One would add oxygen therapy when needed. One would refer for transplantation with steroid therapy failure. Four physicians would first rule out other diseases by open lung biopsy, then initiate cytotoxic drugs.

Of the physicians adding cytotoxic drugs in prednisone failures, six would use cyclophosphamide, two would use azathioprine, and five would use either.

DISCUSSION

This modest survey of pulmonary physicians was illuminating to us. It disclosed, somewhat to our surprise, that the group surveyed approached patients...
with interstitial lung disease in a quite diverse manner. There was no consistency in the diagnostic procedures applied, criteria for the decision to treat, drugs used to treat, or methods for monitoring. Respondents indicated either nonacceptance of literature reports or an experience that did not validate such reports. Indeed, the only consensus views were that better information was needed and that these physicians would be willing to participate in an effort to generate such information. We do not know, of course, whether this modest survey is representative of results that might be found in a more formal, broader survey of pulmonary physicians. Indeed, it appears that the generation of an adequate patient base for any clinical trials may well require collaboration among many pulmonary physicians. A better understanding of current management practices achieved through such a survey would be an invaluable antecedent to the development of a clinical trial protocol.

REFERENCES