New Pulmonary Fibrosis Treatments in Pipeline

BY MARY ELLEN SCHNEIDER
Senior Writer

NEW YORK — Physicians may be getting more options for the treatment of idiopathic pulmonary fibrosis as more therapies come down the research pipeline, Dr. Paul F. Simonelli said at a conference on pulmonary and critical care medicine sponsored by Columbia University.

Among the therapies under evaluation in recently completed or ongoing clinical trials for idiopathic pulmonary fibrosis (IPF), are interferon-gamma-1b, *N*-acetylcysteine, bosentan, etanercept, and imatinib. In addition, there are some molecules, such as pirfenidone, being tested that currently have no other approved uses, said Dr. Simonelli of Columbia University (New York).

The development of new therapies is critical because there are no approved treatments for IPF and the standard approaches are not getting results, he said.

"IPF is a serious disease. It's a debilitating disease, and up to now we've had no effective therapy," Dr. Simonelli said.

The prevalence of the disease is about 83,000 cases in the United States with about 31,000 new cases each year. And the disease has a mortality worse than that of almost any other major disease, except lung cancer. Patients with IPF face a 5-year survivorship of less than 50%, Dr. Simonelli said.

The majority of available data relates to the use of interferon-gamma-1b in IPF. An earlier phase III trial of about 330 patients showed no difference between the drug and placebo for the trial's primary endpoint of progression-free survival (N. Engl. J. Med. 2004;350:125-33). However, a subgroup analysis indicated possible survival benefits with the drug. A second phase III trial looking at survival as the primary end point is underway, and the drug maker InterMune is recruiting patients.

A European study showed promising results for *N*-acetylcysteine in treating IPF. The drug showed improvement in vital capacity and diffusing capacity compared to treatment with prednisone and azathioprine (N. Engl. J. Med. 2005;353:2229-42). But Dr. Simonelli said the results are hard to interpret since the standard of care in Europe is the use of prednisone and azathioprine instead of a true placebo.

The development of new therapies is critical because there are no approved treatments for IPF, and the standard approaches are not getting results.

Another much-discussed possible treatment is pirfenidone. Three trials have been conducted on the drug—an open-label phase II trial in North America, a Japanese trial stopped early because patients on placebo were experiencing severe exacerbations, and a third trial ongoing in Europe. Additional trials of the drug are expected to begin in the United States sometime this year. ■

Variants Key

Metabolic Link *from page 1*

High numbers of lesions also correlated with increases in erythrocyte sedimentation rate and C-reactive protein levels.

A total of 78% patients had the plaque variant of morphea, with the remainder having the guttate variant, idiopathic atrophoderma of Pasini and Pierini, linear scleroderma, and profound scleroderma.

In patients with all variants of morphea, lesions were found on the trunk in 81%, while facial lesions were seen in only eight patients. Overlap syndromes also were reported; eight patients had morphea and lichen sclerosus et atrophicus, and two had morphea with eosinophilic fasciitis.

"Our data also suggest the existence of variant-specific organ involvement in morphea," Dr. Pfeiffer said. Arthralgias were reported by 40% of patients with atrophoderma Pasini and Pierini, while linear scleroderma was associated with the presence of antinuclear antibodies, muscular atrophy, and contractures. Among patients with profound scleroderma, 45% had myalgias and myopathy.

There were no increases in Raynaud symptoms, carpal tunnel syndrome, or lung disorders in patients with any of the variants, she said. ■