

**CORRESPONDENCE**

- 1458 Prevention of Postoperative Nausea and Vomiting  
1459 Airway Obstruction in COPD  
1461 Management of COPD  
1463 Open vs. Laparoscopic Mesh Hernia Repair  
1465 Erythropoietin, Glutamate, and Neuroprotection  
1466 Remission of Lymphoma with Daclizumab

**BOOK REVIEWS**

- 1468 Stem Cell Transplantation for Hematologic Malignancies  
1469 Atlas of Blood Cells: Function and Pathology

**CONTINUING MEDICAL EDUCATION**

- 1473 Treatment of Pulmonary Arterial Hypertension  
1474 Caspofungin versus Liposomal Amphotericin B in Patients with Persistent Fever and Neutropenia  
1475 Footprints

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## Next Week in the Journal

OCTOBER 7, 2004

**50 Years against Polio  
1954–2004**

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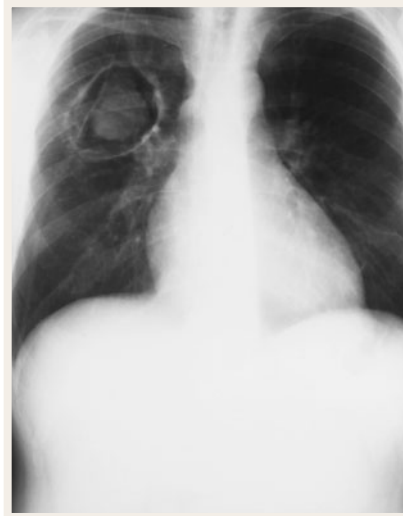


# This Week in the Journal

SEPTEMBER 30, 2004

**ORIGINAL ARTICLE**

### Antifungal Therapy in Patients with Fever and Neutropenia



This multinational, double-blind trial compared caspofungin, an echinocandin, with liposomal amphotericin B as empirical antifungal therapy in 1095 patients with persistent fever and neutropenia. Caspofungin was as efficacious as the standard therapy and was better tolerated, with less nephrotoxicity and fewer drug-related adverse events.

The results of this trial support an effective new option for empirical antifungal therapy in high-risk patients with persistent fever and neutropenia.

SEE P. 1391; EDITORIAL, P. 1445;  
CME, P. 1474

## ORIGINAL ARTICLE

**Pure Red-Cell Aplasia and Epoetin Therapy**

In 2002, the *Journal* published a report of 13 cases of pure red-cell aplasia in patients with renal failure who were receiving erythropoietin (epoetin). From 1998 through 2003, 175 cases were associated with the use of Eprex (epoetin alfa), 11 with the use of Neorecormon (epoetin beta), and 5 with the use of Epogen (epoetin alfa). After the promulgation of new regulatory mandates, the incidence of pure red-cell aplasia associated with epoetin therapy was reduced by more than 80 percent.

Postmarketing surveillance, modifications of manufacturing practices, and changes in the route of administration sharply reduced the incidence of Eprex-associated red-cell aplasia.

SEE P. 1403; PERSPECTIVE, P. 1385

## ORIGINAL ARTICLE

**Autoimmune Lymphoproliferative Syndrome with Somatic *Fas* Mutations**

The autoimmune lymphoproliferative syndrome (ALPS) is a childhood disorder in which lymphadenopathy, splenomegaly, hypergammaglobulinemia, and autoimmunity can be traced to the resistance of lymphocytes to apoptosis. The failure of cell death is usually due to a germ-line mutation of *Fas*, a gene that is essential for apoptosis. This article describes a form of the disease involving a somatic mutation in *Fas*.

These studies highlight the importance of the *Fas* gene in maintaining homeostasis of the lymphoid system.

SEE P. 1409; PERSPECTIVE, P. 1388

## BRIEF REPORT

**Inherited Perforin and *Fas* Mutations in a Patient with Autoimmune Lymphoproliferative Syndrome and Lymphoma**

A patient who inherited a mutation in the *Fas* gene from his father and a mutation in the perforin gene from his mother was found to have autoimmune lymphoproliferative syndrome (ALPS) and a B-cell lymphoma. The mutant *Fas* gene was probably responsible for the development of ALPS, and the mutant perforin gene probably diminished resistance to the development of a B-cell lymphoma.

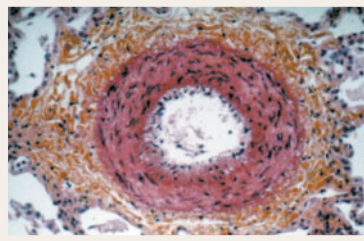
SEE P. 1419; PERSPECTIVE, P. 1388

## DRUG THERAPY

**Treatment of Pulmonary Arterial Hypertension**

Pulmonary arterial hypertension, which is characterized by vascular proliferation and remodeling of small pulmonary vessels, leads to a progressive increase in pulmonary vascular resistance and, ultimately, to right ventricular failure and death. Despite recent major improvements in therapy, no current treatments cure this devastating condition. This article discusses recent progress in developing treatments that prolong patients' lives and improve their quality of life.

SEE P. 1425; CME, P. 1473



## CLINICAL PROBLEM-SOLVING

**Footprints**

A 42-year-old woman with a history of asthma and the irritable bowel syndrome presented with pain and swelling of her feet and ankles that were associated with a rash on her lower extremities.

SEE P. 1438; CME, P. 1475

## CLINICAL IMPLICATIONS OF BASIC RESEARCH

**Chitin and Asthma**

Interleukin-13 has an important influence on the pathophysiology of asthma. A recent study shows that a chitinase may be responsible for the effects of interleukin-13 and is critical to the asthmatic response in a mouse model.

SEE P. 1455

## HEALTH POLICY REPORT

**Linking Physicians' Pay to Quality of Care in the United Kingdom**

In this Health Policy Report, the author describes a major pay-for-performance program that is being implemented in the United Kingdom. Additional government payments to family practitioners will be based on the quality of care they deliver. Family practitioners can earn up to 1050 quality bonus points (expected to be worth more than \$75,000 in gross income) for performing well on a complex set of indicators that measure the quality of clinical care, organization of the practice, and experience of the patients.

SEE P. 1448