



# This Week in the Journal

FEBRUARY 5, 2004

**CORRESPONDENCE**

- 616 Ximelagatran versus Warfarin after Total Knee Replacement
- 618 Ximelagatran for Secondary Prevention of Venous Thromboembolism
- 619 Fondaparinux in Pulmonary Embolism
- 619 First-Trimester Screening for Down's Syndrome
- 622 Effects of Estrogen plus Progestin on Health-Related Quality of Life
- 623 Potts Shunt in Patients with Pulmonary Hypertension
- 624 Tissue Matching before the Era of HLA Typing

**BOOK REVIEWS**

- 625 Goldberger's War: The Life and Work of a Public Health Crusader
- 627 Public Health and the Risk Factor: A History of an Uneven Medical Revolution
- 627 Rockefeller Philanthropy and Modern Biomedicine: International Initiatives from World War I to the Cold War

**CONTINUING MEDICAL EDUCATION**

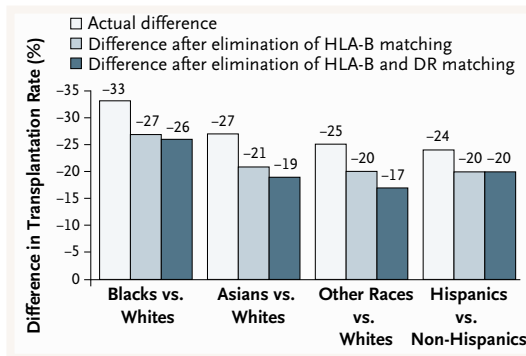
- 633 The Budd–Chiari Syndrome
- 634 Parvovirus B19
- 635 Effect of Eculizumab on Hemolysis and Transfusion Requirements in Patients with Paroxysmal Nocturnal Hemoglobinuria

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**ORIGINAL ARTICLE**

**HLA Matching and Kidney Transplantation in Minority Groups**



In the United States, proportionately more white patients than nonwhite patients who are on waiting lists for transplantation receive a renal transplant. This study compared the rates of transplantation among various racial and ethnic groups with the use of the current allocation policy, a policy in which HLA-B matching was eliminated as a priority, and a policy in which both HLA-B and DR matching were removed as priorities. Removing HLA-B matching as a priority could reduce the existing racial imbalance by increasing the number of nonwhites who receive a transplant, with only a small increase in graft loss.

Racial and ethnic parity in kidney transplantation would be encouraged by changing the current allocation policy.

SEE P. 545; PERSPECTIVE, P. 535

## ORIGINAL ARTICLE

**Eculizumab in Patients with Paroxysmal Nocturnal Hemoglobinuria**

The cause of the hemolytic anemia of paroxysmal nocturnal hemoglobinuria (PNH) is excessive susceptibility of erythrocytes to the lytic effects of the membrane-attack complex of the complement system. Cleavage of C5 initiates assembly of this complex. Eculizumab, a humanized antibody that blocks cleavage of C5, reduces signs of hemolytic anemia and transfusion requirements in patients with PNH.

This open-label study indicates that eculizumab is a safe and effective treatment for PNH. Nevertheless, physicians treating this disease should be cautious, because the long-term effects of eculizumab are unknown.

SEE P. 552; PERSPECTIVE, P. 537

## ORIGINAL ARTICLE

**Association of Cystic Fibrosis with Fatty Acid Metabolism**

Patients with cystic fibrosis have altered levels of plasma fatty acids, but whether these abnormalities reflect inflammation or a primary defect in fatty acid metabolism is not known. In this study, investigators profiled fatty acids in subjects with cystic fibrosis, healthy controls, subjects with inflammatory bowel disease, and subjects with asthma. The ratio of arachidonic to docosahexaenoic acid was increased in subjects with cystic fibrosis, as compared with healthy controls.

Genetic abnormalities of the cystic fibrosis transmembrane conductance regulator are associated with abnormal profiles of tissue and plasma fatty acids.

SEE P. 560; EDITORIAL, P. 605

## BRIEF REPORT

**IGF Deficiency Caused by an IGFALS Mutation**

Insulin-like growth factor I (IGF-I) circulates predominantly as a 150-kD ternary complex that includes the ligand itself, IGF-binding protein 3, and an acid-labile subunit that stabilizes the complex. This report describes a 17-year-old boy with delayed onset and progression of puberty and yet minimal slowing of his linear growth. The boy was found to have an inactivating mutation of the IGF acid-labile subunit (*IGFALS*) gene.

This mutation appears to be capable of causing a marked decrease in the level of circulating IGF-I and delayed puberty.

SEE P. 570

## MECHANISMS OF DISEASE

**Parvovirus B19**

Parvovirus B19 is the cause of fifth disease in children and can trigger transient arthropathy in adults. It can also provoke transient aplastic crises in patients with sickle cell disease or other chronic hemolytic anemias and cause pure red-cell aplasia. This article reviews the features of the virus, the host response to it, and ways of preventing and treating the infection.

SEE P. 586



## CURRENT CONCEPTS

**The Budd–Chiari Syndrome**

Hepatic venous outflow obstruction may develop at the level of the hepatic venules, the large hepatic veins, the inferior vena cava, or the right atrium. Outflow obstruction results in increased hepatic sinusoidal pressure and portal hypertension. This review summarizes the causes of the Budd–Chiari syndrome and current management, including the use of anticoagulant therapy, thrombolytic therapy, transjugular intrahepatic shunts, surgical portosystemic shunts, and liver transplantation.

SEE P. 578

## CLINICAL PROBLEM-SOLVING

**Occam's Razor versus Saint's Triad**

A 60-year-old woman with a history of seronegative rheumatoid arthritis presented to the emergency department with a 10-day history of worsening dyspnea on exertion, nonproductive cough, and fever and a 7-day history of pain in the right leg and buttock.

SEE P. 599

## CLINICAL IMPLICATIONS OF BASIC RESEARCH

**Reducing the Risk of Graft-versus-Host Disease**

A recent study found that including a robust fraction of regulatory T lymphocytes in the inoculum of hematopoietic stem cells reduced the risk of graft-versus-host disease in a mouse model of lymphoma.

SEE P. 614