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Malignant Lymphomas
Gain of chromosome region 18q21 including the MALT1 gene is associated with the activated B-cell-like gene expression subtype and increased BCL2 gene dosage and protein expression in diffuse large B-cell lymphoma
Judith Dierlamm, Eva M. Alurga Penas, Stefan Bendtink, Swen Wessendorf, Hilmar Berger, Michael Hummel, Wolfram Klapper, Didol Lensze, Andreas Rosenwald, Eugenia Haralambieva, German Ott, Sergio B. Cogliatti, Peter Möller, Carsten Schwaneen, Harald Stein, Markus Leffler, Rainer Spang, Lorenz Trümper, and Reiner Siebert for the Deutsche Krebshilfe Network Project “Molecular Mechanisms in Malignant Lymphomas”

Diffuse large B-cell lymphoma is the most common lymphoma worldwide. Recent studies of gene expression profiling have identified subgroups of this lymphoid neoplasm...
with different prognosis. This study suggests that a gain of chromosome 18q21 including the MALT1 gene may  involve an unfavorable prognosis. See related perspective article on page 641.

### Lymphoproliferative Disorders

#### Selective influences in the expressed immunoglobulin heavy and light chain gene repertoire in hairy cell leukemia

Francesco Forconi, Elisa Sozzi, Davide Rossi, Surinder S. Sahota, Teresa Amato, Donatella Raspadori, Livio Trentin, Lorenzo Leoncini, Gianluca Gaidano, and Francesco Lauria

Hairy cell leukemia is a rare, chronic B-cell neoplasm characterized by leukemic hairy cells. This immunogenetic analysis of the expressed immunoglobulin heavy and light chain gene repertoire suggests that immunoglobulin gene selection may play an important role in the pathogenesis of this neoplasm.

### Thrombocytosis

#### A de novo splice donor mutation in the thrombopoietin gene causes hereditary thrombocytopenia in a Polish family

Kun Liu, Robert Kralovics, Zbigniew Rudzki, Barbara Grabowska, Andreas S. Buser, Damla Okaydu, Heinz Gislinger, Ralph Tiedt, Patricia Frank, Krzysztof Okon, Anthonie P.C. van der Maas, and Radek C. Skoda

Familial thrombocytosis may be associated with gain-of-function mutations in the thrombopoietin (THPO) gene, or an activating germline mutation of MPL, the gene encoding thrombopoietin receptor. This study of a large family with thrombocytosis associated with a germline THPO mutation illustrates the hematologic and clinical features of this rare condition. See related article on page 646.

### Disorders of Hemostasis

#### Simultaneous genotyping of coagulation factor XI type II and type III mutations by multiplex real-time polymerase chain reaction to determine their prevalence in healthy and factor XI-deficient Italians

Giorgia Zadra, Rosanna Asselta, Maria Luisa Trenchini, Giancarlo Castaman, Uri Seligson, Pier Mannuccio Mannucci, and Stefano Duga

Factor XI deficiency is a rare autosomal recessive coagulopathy worldwide, although relatively common in Ashkenazi Jews. This study describes the presence of the F11 gene mutations in the Italian population and confirms their Jewish origin.

### Disorders of Hemostasis

#### The Italian AICE-Genetics hemophilia A database: results and correlation with clinical phenotype

Maurizio Maragoulone, Giancarlo Castaman, Massimo Morfaji, Angela Rocino, Elena Santagostino, Giuseppe Tagariello, Anna R. Tagliaferri, Ezio Zanon, Maria P. Bicocchi, Giuseppe Castaldo, Flora Peviani, Rosa Santacroce, Francesca Torricelli, Elisa Grandone, Pier Mannuccio Mannucci, and the AICE-Genetics Study Group

This study reports a wide spectrum of factor 8 mutations in the large Italian database. Findings of the study indicate that the type of mutations is a strong predictor of the clinical phenotype.

### Thrombosis

#### AB0 blood group and risk of venous or arterial thrombosis in carriers of factor V Leiden or prothrombin G20210A polymorphisms

Antonia Miñano, Adriana Ordóñez, Francisco España, José R. González-Porras, Ramón Lecumberri, Jordi Fontalberta, Pilar Llamas, Francisco Marín, Amparo Estellés, Ignacio Alberca, Vicente Vicente, and Javier Corral

Several studies have shown an effect of AB0 blood group on hemostasis. Findings of this study indicate that non-00 group increases the risk and severity of venous thrombosis in carriers of prothrombotic polymorphisms. See related perspective article on page 649.

### Stem Cell Transplantation

#### Follow-up of healthy donors receiving granulocyte colony-stimulating factor for peripheral blood progenitor cell mobilization and collection. Results of the Spanish Donor Registry

Javier de la Rubia, Felipe de Arriba, Cristina Arbona, María J. Pascual, Concha Zamora, Andrés Insunza, Dorleta Martínez, Carmen Paniagua, Miguel A. Díaz, and Miguel A. Sans

Information about the long-term follow-up and safety of granulocyte colony-stimulating factor administration to healthy donors is limited. Findings of this study indicate that clinical side effects are generally mild, and that development of secondary hematologic malignancy is unlikely.

### Decision Making and Problem Solving

#### Italian Society of Hematology practice guidelines for the management of iron overload in thalassemia major and related disorders

Emanuele Angelucci, Giovanni Barosi, Clara Camaschella, Maria Domenica Cappellini, Mario Cazzola, Renzo Galanello, Monia Marchetti, Antonio Piga, and Sante Tura

Three iron chelators are currently available for treatment of transfusion iron overload: deferoxamine, deferasirox, and deferasirox. This article reports the Italian Society of Hematology practice guidelines for the management of iron overload in thalassemia major and related disorders.
May 2008

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Brief Reports

Iron Metabolism

753 Deficiency of heme-regulated eIF2α kinase decreases hepcidin expression and splenic iron in HFE−/− mice
Sijin Liu, Rajasekhar N.V.S. Suragani, Anping Han, Wanting Zhao, Nancy C. Andrews, and Jane-Jane Chen

Heme-regulated eIF2α balances heme and globin production by controlling globin synthesis. Its deficiency affects the phenotype of HFE−/− mice by further decreasing hepcidin expression.

Sickle Cell Disease

757 Sickle cell disease-related organ damage occurs irrespective of pain rate.
Implications for clinical practice
Eduard J. van Beers, Charlotte F.J. van Tuijn, Melvin R. Mac Gillavry, Anna van der Giessen, John-John B. Schnog, and Bart J. Biemond, on behalf of the CURAMA study group

This study shows that clinically relevant forms of organ damage occur irrespective of the frequency of painful crises in adults with sickle cell disease.

Anemia of Renal Failure

761 Anemia management in patients on peritoneal dialysis: efficacy and safety of epoetin δ
Bruce S. Spinowitz

In this clinical trial, subcutaneously administered epoetin δ was effective and well tolerated for treatment of anemia in patients on peritoneal dialysis.

Chronic Myeloid Leukemia

765 Phospho-CRKL monitoring for the assessment of BCR-ABL activity in imatinib resistant chronic myeloid leukemia or Ph+ acute lymphoblastic leukemia patients treated with nilotinib
Paul La Rosée, Susanne Holm-Eriksen, Heiko Konig, Nicolai Härtel, Thomas Ernst, Julia Debatin, Martin C. Mueller, Philipp Erlen, Anja Binckebanck, Lydia Wanderle, Yaqing Shou, Margaret Dugan, Ruediger Hethlmann, Oliver G. Ottmann, and Andreas Hochhaus

Findings of this study suggest that monitoring the actual BCR-ABL inhibition in nilotinib treated patients may be useful for establishing effective dosing and for detecting resistance against the drug.

Chronic Myeloid Leukemia

770 Front-line treatment of Philadelphia positive chronic myeloid leukemia with imatinib and interferon-α: 5-year outcome
Francesca Palandrini, Iatira Iacobucci, Fausto Castagnetti, Nicoletta Testori, Angela Poerio, Marilina Amabile, Massimo Breccia, Tamara Intermesoli, Francesco Iuliano, Giovanni Rege-Cambrin,

Thalassemia Syndrome

784 A cross-sectional magnetic resonance imaging assessment of organ specific hemosiderosis in 180 thalassemia major patients in Hong Kong
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787 The Janus kinase 2 (JAK2) V617F mutation in Chinese patients with chronic myeloproliferative disorders
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Acute Myeloid Leukemia

785 Cytoplasmic mutated nucleophosmin is stable in primary leukemic cells and in a xenotransplant model of NPMc+ acute myeloid leukemia in SCID mice
Brunangelo Falini, Maria Paola Martelli, Cristina Mecucci, Arcangelo Loso, Niccolò Bolli, Barbara Bigerna, Alessandra Puciarini, Stefano Pileri, Giovanna Meloni, Massimo F. Martelli, Torsten Haferlach, and Susanne Schnittger

Findings of this study indicate that NPM1 mutations are stable in patients with acute myeloid leukemia, providing a rationale for monitoring of minimal residual disease.

Malignant Lymphomas

780 Clinical and demographic characteristics of Epstein-Barr virus-associated childhood Burkitt's lymphoma in Southeastern Brazil: epidemiological insights from an intermediate risk region
Rocio Hassan, Claudete Esteves Klumb, Fabrício E. Felisbino, Deisy M. Guretti, Lidia R. White, Claudio Gustavo Stefano, Mario Henrique M. Barros, Héctor N. Seuánez, and Ilana R. Zalcberg

This report describes clinical and demographic characteristics of Epstein-Barr virus-associated childhood Burkitt's lymphoma in Southeastern Brazil.

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