Cover Figure

Congo red staining and analysis under polarized light showing the diagnostic green birefringence of amyloid deposits. This illustration is taken from the perspective article by Drs. Palladini and Merlini on page 1044.

Editorials and Perspectives

1039 The hereditary stomatocytoses
Joanna F. Flatt and Lesley J. Bruce

Hereditary stomatocytosis describes a wide spectrum of autosomal dominantly inherited hemolytic disorders in which the basal red cell membrane cation permeability is increased. In this perspective article, Drs. Flatt and Bruce summarize our current knowledge in the field. See related article on page 1049.

1041 Flow cytometry immunophenotyping for diagnosis of myelodysplastic syndrome
Mario Cazzola

In this perspective article, Dr. Cazzola examines standard and novel tools for the diagnosis of myelodysplastic syndrome. Flow cytometry immunophenotyping may provide complementary information in the diagnostic approach to a patient with this condition. See related articles on pages 1066, 1124 and 1160.

1044 Current treatment of AL amyloidosis
Giovanni Palladini and Giampaolo Merlini

Several effective chemotherapy regimens have been developed during the last decade significantly improving the outlook of patients with AL amyloidosis. Drs. Palladini and Merlini describe our current knowledge in the field and examine future perspectives. See related article on page 1049.

Original Articles

Red Cell Disorders

1049 A novel erythroid anion exchange variant (Gly796Arg) of hereditary stomatocytosis associated with dyserythropoiesis
Achille Iolascon, Luigia De Falco, Franck Borgese, Maria Rosaria Esposito, Rosa Anna Avisati, Pietro Izzo, Carmelo Piscopo, Helene Guizouarn, Andrea Biondani, Antonella Pantaleo, and Lucia De Franceschi

Stomatocytoses are a group of inherited autosomal dominant hemolytic anemias and include overhydrated hereditary stomatocytosis, dehydrated hereditary stomatocytosis, hereditary crythrocytosis and familial pseudohyper-...
Myeloproliferative Neoplasms

1085 The fusion proteins TEL-PDGFβ and FIP1L1-PDGFRα escape ubiquitination and degradation
Federica Toffalini, Anders Kallin, Peter Vandenbergh, Pascal Pierre, Lucienne Michaux, Jan Cools, and Jean-Baptiste Demoulin

Upon growth factor-induced activation, receptor tyrosine kinases such as the PDGF and FGF receptors are targeted for lysosomal degradation via a mechanism that involves ubiquitination of receptor lysines. In this study, it is shown that constitutively active oncogenic fusion proteins that contain PDGF or FGF receptor moieties, caused by specific chromosomal translocations in chronic myeloid neoplasms, escape this negative regulatory mechanism.

Amyloidosis

1094 Histological regression of amyloid in AL amyloidosis is exclusively seen after normalization of serum free light chain
Ingrid I. van Gameren, Martin H. van Rijswijk, Johan Bijzet, Edo Vellenga, and Bouke P. Hazenberg

Amyloidosis is thought to be a dynamic process of deposition and resolution. This implies that after elimination of the precursor protein, amyloid deposits in organs might resolve in the course of time resulting in improvement of both organ function and clinical performance. The findings of this study indicate that achievement of complete response of amyloidogenic free light chain following chemotherapy is associated with a significant reduction in amyloid deposition in fat tissue. See related perspective article on page 1044.

Stem Cell Transplantation

1101 Timing and severity of community acquired respiratory virus infections after myeloablative versus non-myeloablative hematopoietic stem cell transplantation
Joshua T. Schiffer, Kate Kirby, Brenda Sandmaier, Rainer Storb, Lawrence Corey, and Michael Boeckh

Respiratory virus infections are important causes of morbidity and mortality after hematopoietic cell transplantation. Their clinical course can be severe with progression to lower respiratory tract infection, co-infection with serious pulmonary co-pathogens, and high mortality. The findings of this retrospective cohort study indicate that viral lower respiratory tract infection during the first 100 days after hematopoietic cell transplantation was less common among patients receiving non-myeloablative conditioning regimens than in those receiving myeloablative conditioning, despite a similar overall rate of acquisition.

Decision Making and Problem Solving

1109 Chronic inflammatory disease, lymphoid tissue neogenesis and extranodal marginal zone B cell lymphomas
Richard J. Bendele, Febe van Maldegem, and Carael J.M. van Noesel

Chronic autoimmune or pathogen-induced immune reactions resulting in lymphoid neogenesis are associated with development of malignant lymphomas, mostly extranodal marginal zone B-cell lymphomas. This review article examines the role of chronic inflammatory responses and the molecular mechanisms involved in the development and progression of extranodal marginal zone B-cell lymphomas.

Myelodysplastic Syndromes

1124 Standardization of flow cytometry in myelodysplastic syndromes: report from the first European LeukemiaNet working conference on flow cytometry in myelodysplastic syndromes

This article describes the results of the first European LeukemiaNet working conference on flow cytometry in myelodysplastic syndromes. This report is a very comprehensive analysis of the topic, and provides detailed information on what is currently known in the field. See related perspective article on page 1044.

Acute Leukemia

1135 How and why minimal residual disease studies are necessary in leukemia: a review from WP10 and WP12 of the European LeukaemiaNet
Marie C. Béné and Jaspal S. Kaeda

Disease reappears in the majority of leukemia patients who enter remission. For many years investigators have focused on detecting the "minimal residual disease" ultimately responsible for these relapses. Two primary methods, one based on polymerase chain reaction technology, the other on flow cytometry, are in increasing use. This paper describes the application of such methodologies not only to chronic myeloid leukemia, childhood acute lymphoblastic leukemia, and acute promyelocytic leukemia but to other types of leukemia.
**Brief Reports**

**Hematopoietic Stem Cells**

Mesenchymal stem cells efficiently inhibit the proinflammatory properties of 6-sulfo LacNAc dendritic cells

Rebekka Wehner, Diana Wehrum, Martin Bornhäuser, Senming Zhao, Knut Schäkel, Michael P. Bachmann, Uwe Platzbecker, Gerhard Eltunanger, E. Peter Rieber, and Marc Schmitz

Mesenchymal stem cells (MSC) exert modulatory effects on the immune system and may have a role in the treatment of steroid-refractory graft-versus-host disease. Here it is shown that MSC inhibit the maturation of a subpopulation of human dendritic cells called slanDC, thereby impairing their ability to produce proinflammatory cytokines and to stimulate the proliferation of CD4+ and CD8+ T lymphocytes.

**Thalassemia Syndromes**

Severe intrauterine anaemia: a new form of \(\epsilon\gamma\delta\beta\) thalassemia presenting in utero in a Norwegian family

Anne Brantberg, Sturla H. Eik-Nes, Nigel Roberts, Chris Fisher, and William G. Wood

This report describes a type of \(\epsilon\gamma\delta\beta\)-thalassemia due to a rare private deletion, detected early and successfully treated with intrauterine blood transfusions.

**Myelodysplastic Syndromes**

Reduced CD38 expression on CD34+ cells as a diagnostic test in myelodysplastic syndromes

Nicolas Goardon, Emmanouil Nikolousis, Alexander Sternberg, Wai-Kit Chu, Charles Craddock, Peter Richardson, Richard Benson, Mark Drayson, Graham Stauden, Parsh Vyas, and Sylvie Freeman

This report indicates that a reduced mean fluorescence intensity of CD38 expression on CD34+ cells can be used as a surrogate marker for abnormalities in the CD34+ compartment of patients with myelodysplastic syndrome. See related perspective article on page 104.

**Acute Lymphoblastic Leukemia**

Heterogeneous breakpoints in patients with acute lymphoblastic leukemia and the dic(9;20)(p11-13;q11) show recurrent involvement of genes at 20q11.21

Qian An, Sarah L. Wright, Anthony V. Moorman, Helen Parker, Mike Griffiths, Fiona M. Ross, Teresa Davies, Christine J. Harrison, and Jon C. Strefford

Dentric chromosomes are rare in acute lymphoblastic leukemia, dic(9;20) being a recurrent aberration. This study provides insight into the breakpoint complexity underlying dentric chromosomal formation in acute lymphoblastic leukemia and highlights putative target gene loci.

**Errata Corrige**

High INDO (indoleamine 2,3-dioxygenase) mRNA level in blasts of acute myeloid leukemic patients predicts poor clinical outcome. Haematologica 2008; 93:1894-1898.


Late relapse of acute myeloid leukemia with mutated NPM1 after eight years: evidence of NPM1 mutation stability. Haematologica 2009; 94:298-300.

Meloni G, Mancini M, Gianfelici V, Martelli MP, Foa R, Falini B

1183 Haematologica 2009; 94[supplement 2]

1183 Definition, diagnosis and treatment of immune thrombocytopenic purpura. Haematologica 2009; 94:759-762

George JN

1184 Regulation of serum hepcidin levels in sickle cell disease. Haematologica 2009;94:885-887

Kroot JJ, Laarakkers CM, Kemna EH, Biemond BJ, Swinkels DW.

Continuing Medical Education

Community-acquired respiratory virus infections after myeloablative versus non-myeloablative conditioning for transplantation

Histological regression of amyloid in AL amyloidosis

Pathogenesis of extranodal marginal zone B-cell lymphomas

Diagnostic utility of flow cytometry in myelodysplastic syndromes