HÉMA-QUÉBEC

HÉMA-VİGİE…always on the lookout!

A Monthly Newsletter Summarizing Important Advances in Transfusion

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An Immunotherapeutic Approach to Treating Prion Diseases

Transmissible Spongiform Encephalopathies (TSEs), or prion diseases, refer to a group of neurodegenerative pathologies caused by intracerebral accumulation of an aberrant form (PrP^{Sc}) of the prion protein normally present in its natural conformation (PrP^{C}) in several tissues. Sheep scrapie, mad-cow disease, and new variant Creutzfeld-Jakob disease (nvCJD) in humans are well-known examples of such illnesses. One of the unique properties of the pathogenic PrP^{Sc} protein is its infectious behaviour. The possible link between mad-cow disease and nvCJD, and the hypothetical risk of nvCJD transmission by blood transfusion, prompts blood collection agencies to keep track of scientific advances in the diagnosis and treatment of prion diseases. At this time, these diseases are invariably fatal, as no effective therapies are available. In the October 5 issue of *Science*, Adriano Aguzzi (University Hospital Zurich, Zurich, Switzerland) and colleagues report on experimental results suggesting that continued expression of an anti-prion antibody renders mice resistant to experimental infection with a pathogenic strain of prion protein.

Heppner, F. L. et al., ***Prevention of Scrapie Pathogenesis by Transgenic Expression of Anti-Prion Protein Antibodies***, Science 294 (5540): 178-182 (2001).

Identification of a Susceptibility Gene for Hereditary Thrombocytopenia

Thrombotic Thrombocytopenia purpura (TTP) is a coagulation disorder which manifests itself by excessive aggregation of blood platelets, leading to serious thrombotic episodes in peripheral tissues, especially the brain and kidneys. An unusually high frequency of thrombotic incidents in a few rare families suggests that some TTPs are genetically inherited. David Ginsburg (Howard Hughes Medical Institute, University of Michigan Medical Center, Ann Arbor, MI, USA) and colleagues report, in the 4 October issue of *Nature*, the identification of the gene responsible for hereditary TTP. The protein encoded by the gene is a plasma enzyme whose function is to cleave von Willebrand multimers, which are protein complexes also involved in blood coagulation. These results shed new light on the molecular mechanisms of blood coagulation, and mark the starting point towards the development of new therapies for these patients.

Levy, G. G. et al., ***Mutations in a Member of the ADAMTS Gene Family Cause Thrombotic Thrombocytopenic Purpura***, Nature 413 (6855): 488-494 (2001).

Muscle, as a source of blood cells

Stem cell research continues to arouse a lot of interest within the research community, as well as in the media and among the public in general. Hiroshi Kawada and Makio Ogawa, from the Medical University of South Carolina (Charleston, SC, USA), document the presence of hematopoietic cells in the skeletal muscles of the mouse. Their experimental results, reported in the 1 October issue of *Blood*, suggest that blood cell precursors can possibly be found in several nonhematopoietic tissues throughout the body.

Kawada, H and Ogawa, M., ***Bone Marrow Origin of Hematopoietic Progenitors and Stem Cells in Murine Muscle***, Blood 98 (7): 2008-2013 (2001).

Evaluation of Seven CMV Detection Tests for Blood Screening

Newborns and transplant recipients comprise a significant fraction of patients requiring blood transfusions. However, the immunosuppressed state of these patients makes them particularly vulnerable to cytomegalovirus (CMV) infection. John D. Roback (Emory University, Atlanta, GA, USA) and colleagues undertook the evaluation of seven CMV tests designed for systematic blood screening. Their experimental results, published in the October issue of *Transfusion*, indicate that three of the seven tests demonstrate a sufficient level of sensitivity and specificity for systematic CMV screening among blood donors.

Roback, J. D. et al., ***Multicenter Evaluation of PCR Methods for Detecting CMV DNA in Blood Donors***, Transfusion 41 (10): 1249-1257 (2001).