

Contents

Johann Lukas Schoenlein Prize 2006 I. SCHARRER	1
I. Epidemiology	
HIV Infection and Causes of Death in Patients with Hemophilia in Germany (Year 2005/2006 Survey) W. SCHRAMM and H. KREBS on behalf of the participating German Hemophilia centers	5
Hemophilia Registry of the Medical Committee of the Swiss Hemophilia Association – Update and Annual Survey 2006 S. HARTMANN and K. LOCHER	17
II. Treatment of Inhibitors in Hemophiliacs	
Detection of Factor VIII-Specific Memory B Cells in Patients with Hemophilia A and Factor VIII Inhibitors I. LANG, J. WINDYGA, A. KLUKOWSKA, J. ILAS, H. P. SCHWARZ, and B. M. REIPERT	25
Novel Strategies for FVIII: Half-life Prolongation P. L. TURECEK, J. SIEKMANN, K. VARADI, H. P. MATTHIESSEN, A. WEBER, H. GRITSCH, F. SCHEIFLINGER, E.-M. MUCHITSCH, and H. P. SCHWARZ	37
III. Hemophilic Arthropathy and Synovitis	
Predictive Parameters of Fitness in Hemophilic Children A. SEUSER, P. BÖHM, A. KURME, and K. KURNIK	41
Hemophilia and Exercise Project (HEP) Conception and Contents of a “Programmed Sports Therapy” for Hemophilic Patients M. HERBSLEB, CH. PUTA, and TH. HILBERG	45
Results of German Hemophiliacs Self-Assessment of Health and Social Status: With Emphasis on Orthopedic Function W. KALNINS, and W. MONDORF on behalf of DHG = German Hemophilia Society	60
IV. Relevant Hemophilia Treatment 2006	
Developing a German Hemophilia Register B. HASCHBERGER, J. HESSE, M. HEIDEN, and R. SEITZ	67

Acquired Glanzmann Thrombasthenia in a Patient with Myelodysplastic Syndrome A. TRUMMER, A. TIEDE, and R. EISERT	184
Case Report of an Acquired Inhibitor Against Factor XIII R. KLAMROTH, S. WEIHER, M. KUHLMANN, S. GOTTSTEIN, and J. OLDENBURG	187
Elbow Endoprosthesis in Hemophilic Arthropathy R. SCHOLZ, A. SIEGEMUND, and U. SCHOLZ.	190
Pseudohomozygous APC Resistance Report on Two Patients and a Novel Mutation in the Factor V Gene B. MAAK, L. KOCHHAN, P. HEUCHEL, and F. H. HERRMANN	193
Pseudo-Homozygous APC Resistance Due to Coinheritance of Heterozygous Factor V – R506Q and Type I Deficiency Associated with Thrombosis A. PAVLOVA, R. LORETH, D. DELEV, and J. OLDENBURG	198
Three Cases of In-stent Thrombosis in Patients with Clopidogrel Non-response A. CALATZIS, M. SPANNAGL, F. THEISEN, and V. KLAUSS	201
 <i>Vllc) Diagnostics</i>	
Thrombin Generation in Pregnancy A. ROSENKRANZ, M. HIDEN, B. LESCHNIK, E. WEISS, R. HOFER, D. SCHLEMBACH, U. LANG, and W. MUNTEAN.	205
Thrombin Generation – Comparison of ROTEM and Technothrombin TGA A. SIEGEMUND, S. SAILE, T. SIEGEMUND, U. SCHOLZ, and R. SCHOBESS	212
Multiple Electrode Aggregometry in Patients on Chronic Aspirin and/or Clopidogrel Treatment A. CALATZIS, M. SPANNAGL, and F. THEISEN.	217
Endogenous Thrombin Potential in Women with Recurrent Miscarriage and/or Repeated In-Vitro-Fertilization Failure W.-M. HALBMAYER, W. FEICHTINGER, C. KINDERMANN, M. RANKINE, H. STROHMER, A. OBRUCA, A. SIEGEMUND, and M. BÖHM	218
One Year TTP Diagnosis in the Central Laboratory of the University of Mainz TH. VIGH, I. SCHARRER, D. PEETZ, and K.J. LACKNER	221
 <i>Vll d) Miscellaneous</i>	
Venous Thromboembolism – A Metabolic Disease? C. AY, TH. TENGLER, R. SIMANEK, R. VORMITTAG, TH. VUKOVICH, and I. PABINGER	229
Structural Investigation of Two Novel Mutations in Coagulation Factor V by Molecular Modeling D. DELEV, A. PAVLOVA, E. SEIFRIED, and J. OLDENBURG	231
High Levels of Human Factor IX Transgene Expression in Mice Increase Blood Procoagulant Activity M. A. SROUR, J. GRUPP, TH. ALBERT, J. OLDENBURG, H. FECHNER, R. FIMMERS, and R. SCHWAAB	234
Factor VII Deficiency: Clinical Manifestation and Molecular Genetics of 718 Subjects with FVII Gene Mutations F. H. HERRMANN, K. WULFF, G. AUERSWALD, J. ASTERMARK, A. BATOROVA, W. KREUZ, H. POLLMANN, A. RUIZ-SAEZ, L. SALAZAR-SANCHEZ, S. SCHULMAN, and International Study Group	238

Therapeutical Options for Congenital FVII Deficiency – The HK 7 Project of the International Greifswald Registry of the Congenital FVII Deficiency (GR-HK-7) F. H. HERRMANN, G. AUERSWALD, J. INGERSLEV, CH. SCHUBERT, K. ERDLNBRUCH, F. BOTHA, R. EISERT, A. GERHARDT, B. NIEMANN, E. SCHOMERUS, O. ANDERS, D. FRANKE, M. GIRISCH, A. HUTH-KÜHNE, G. JIMENEZ-CRUZ, R. M. LORETH, L. SALAZAR-SANCHEZ, A. SIEGEMUND, C. WERMES, K. ZWIAUER, and K. WULFF	246
Characterization of a Mutation in Exon 1 of the FVII Gene – a Case of RNA Editing? W. SCHRÖDER, K. WULFF, R. TECH, G. AUERSWALD, S. BECKER, and F. H. HERRMANN	251
Analysis of the Biological Importance of the <i>VKORC1</i> Gene via Knock-Out Mice G. SPOHN, TH. WUNDERLICH, A. KLEINRIDDS, CH. GEISEN, E. SEIFRIED, and J. OLDENBURG.	255
<i>In Vivo</i> -Assay for Warfarin-Resistance by Coexpression of <i>VKORC1</i> and Vitamin K-Dependent Coagulation Factors A. FREGIN, S. ROST, C.R. MÜLLER, and J. OLDENBURG	258
Functional Promoter Polymorphism in the <i>VKORC1</i> Gene is no Major Genetic Determinant for Vitamin K Dependent Coagulation Factor Activity M. MARINOVA, PH. WESTHOFEN, M. WATZKA, B. PÖTZSCH, and J. OLDENBURG	260
Influence of <i>VKORC1</i> Haplotypes on Cardiovascular Disease M. WATZKA, A. NEBEL, N. E. EL MOKHTARI, B. IVANDIC, J. MÜLLER, ST. SCHREIBER, and J. OLDENBURG.	263
Creation and Characterization of a Knock-Out Mouse for the <i>VKORC1LI</i> -Gene M. HÜNERBERG, G. SPOHN, C. R. MÜLLER, and J. OLDENBURG	265
Novel Variants in <i>VKORC1</i> – the Target Protein of Coumarin-Type Anticoagulants – in Rodents from Warfarin-Resistance Areas in Europe, Asia and America S. ROST, H.-J. PELZ, A. FREGIN, M. HÜNERBERG, C. R. MUELLER, and J. OLDENBURG.	268
Recombinant Expression and 3D-Modelling of C1-Inhibitor Mutants T. FÖRSTER, C.R. MÜLLER, and J. OLDENBURG.	270
Five Novel Mutations in F13B Gene Resulting in Mild FXIII Deficiency V. IVASKEVICIUS, H. ROTT, H. TROBISCH, S. HALIMEH, I. SCHARRER, M. KRAUSE, E. SEIFRIED, and J. OLDENBURG	272
Idiopathic Thrombocytopenic Purpura in Children: Predictive Factors for the Outcome M. SERBAN, S. ARGHIRESCU, D. MIHAILOV, A. OPRISONI, M. BATANEANT, L. STANA, M. MATSIRIS, K. SCHRAMM, and W. SCHRAMM.	274
Preoperative DDAVP-Testing in Patients with von Willebrand Disease H.-H. WOLF, K. JORDAN, and A. FRÜHAUF	280