



FINAL PROGRAMME

7TH BARI INTERNATIONAL CONFERENCE

May 21-23 2011

Pugnochiuso Congress Centre
Vieste del Gargano, Foggia, Italy



Welcome

We are pleased to welcome you to the 7th Bari International Conference (BIC), in Pugnochiuso, on the Italian Adriatic Sea coast close to Vieste del Gargano (Foggia).

This meeting has an established tradition, being now at its 7th edition and is quite popular as a small meeting on relative narrow topics (factor VIII, factor IX, rare bleeding disorders, von Willebrand disease, ADAMTS13 and thrombotic microangiopathies). It is jointly organized by the Angelo Bianchi Bonomi Hemophilia and Thrombosis Center of Milan (P.M. Mannucci), the Hemophilia Center of Bari (N. Ciavarella) and the Arteriosclerosis and Thrombosis Centre of La Jolla Scripps Clinic (Z. Ruggeri). The meeting takes place every 3 years, the last time was in May 2008. The venue is included in a natural park located quite far away from the main towns. We hope that this will encourage speakers and participants to remain at the congress venue place for the whole duration of the meeting and enjoy the opportunity of socializing with their international colleagues.

Scientific topics. The meeting runs through 3 full days, with participants arriving on the evening of Friday, May 20th, and departing on Tuesday 24th with the actual sessions from Saturday morning throughout Monday evening (departures on Tuesday 24th May).

We will encourage prospective speakers to present their most recent original data, as they did in the past.

We are also pleased to give room to participants other than those invited, because each day there will be a display of posters.

Oral Communications and poster discussions are scheduled on the last day, Monday 23rd, and will be followed by discussion. In addition, all peer-reviewed accepted abstracts are published in a book of abstracts as a supplement to Blood Transfusion.

Thanks all participants and sponsors who have contributed to the success of this meeting.

Professor PM Mannucci
Chairman

| | | |
|--|------------------------------|----|
| | Committee and Congress Venue | 6 |
| | Congress General Information | 7 |
| | Sponsors | 8 |
| | Floor plan | 9 |
| | Programme | 10 |
| | Speakers | 16 |
| | Notes | 21 |

Venue

Pugnochiuso Congress Centre
Vieste del Gargano, Foggia, Italy

Congress Chairman

Pier Mannuccio Mannucci
Scientific Director of IRCCS Cà Granda
Foundation Ospedale Maggiore Policlinico of
Milan, and President of the Angelo Bianchi
Bonomi Hemophilia Center of Milan, Italy

Scientific Committee

Nicola Ciavarella, Bari, Italy
Zaverio M. Ruggeri, La Jolla, CA, USA

Augusto B. Federici, **Alessandro Gringeri**
Elisa M. Mancuso, **Flora Peyvandi**
University of Milan, Italy

Organizing Secretariat

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Registration and admission

All pre-registered participants are kindly requested to collect their badge and congress kit at the Registration Desk located at the check-in area of Hotel del Faro on Friday 20th and at the Congress Centres in the following days.

Admission to the Scientific and Satellite sessions is reserved only to registered delegates with a regular badge. Please remember to wear your name badge at all times during the Congress.

Congress kit

The Congress kit contains:

- CME educational material
- Programme of the Conference
- Certificate of attendance
- Quality assessment questionnaire
- Eletronic badge
- Book of Abstract

CME (Continuing Medical Education)

European CME: The programme of the 7th Bari International Conference has been reviewed and approved for CME accreditation by the European Hematology Association CME Unit. Each physician should only claim credits for the time that she/he actually spent in the educational activity. Please follow the instructions given in the forms included in your congress kit to be able to claim credits online at the EHA website. European CME credits are reserved to MD in haematology.

Italian CME: The event has been filed with numbers 4577-11012517 and 45577-11012517 for the following specialties: internal medicine, haematology, orthopaedics, pediatrics and biology. The number of credits attributed to this event are 14.

Recording

Any form of duplication (including but not limited to audio/video recording, audio taping, digital taping, photography etc..) of sessions including Poster Presentations is prohibited without the expressed consent of the Congress Organizers.

Food and beverage

Breakfasts and dinners are included in the hotel accommodation package.

Daily coffee breaks and box lunches for those attending lunch symposia are included in the registration fee. Those who do not wish to take part to the lunch sessions may book their lunches upon registration. Vouchers can be purchased at the registration desk at all times but booking is recommended. Lunch will be served at the Congress Venue Restaurant Le Piramidi.

Language

English is the official Congress language.

WE THANK ALL SPONSORS WHO CONTRIBUTED WITH UNRESTRICTED EDUCATIONAL GRANTS TO THIS MEETING

Baxter

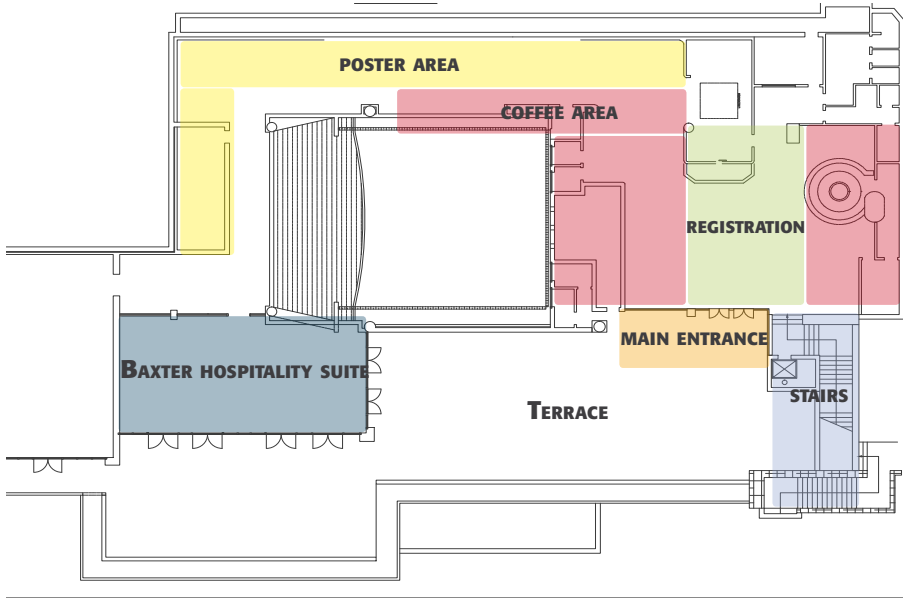
Pfizer *Speciality Care*



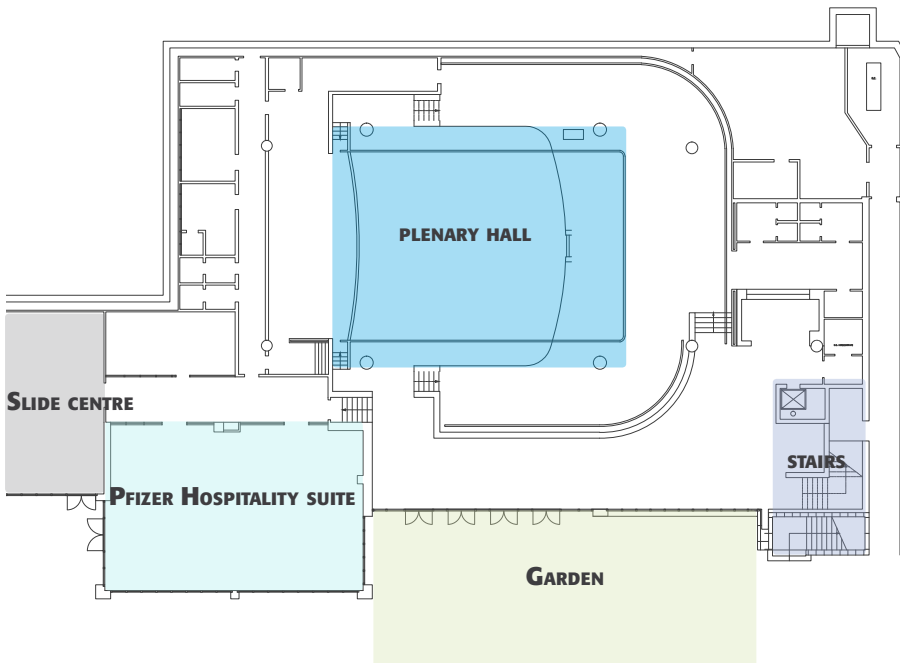
CSL Behring



Congress Centre



UPPER FLOOR



BASEMENT

Saturday, 21st May

8:30-10:30

SCIENTIFIC SESSION 1

Chairs: Pier Mannuccio Mannucci, Italy; Nicola Ciavarella, Italy

- Fibrinogen - a universal haemostatic agent. [Benny Sørensen](#), UK
- How safe are current clotting factor concentrates? [Mike Makris](#), UK
- Moderate haemophilia: patients at risk of undertreatment. [Kathlyn Fischer](#), the Netherlands
- Acquired haemophilia: survey and treatment of 501 cases. [Francesco Baudo](#), Italy

10:30-11:00 break

11:00-13:00

SCIENTIFIC SESSION 2

Chairs: Bernhard Lämmle, Switzerland; Samuel Machin, UK

- ADAMTS13 in health and disease. [Karen Vanhoorelbeke](#), Belgium
- ADAMTS13 in TTP. [Johanna A. Kremer-Hovinga](#), Switzerland
- ADAMTS13, thrombotic microangiopathies and pregnancy: the French experience. [Alain Stepanian](#), France
- Risk factors and triggering factors in acquired idiopathic TTP. [Paul Coppo](#), France

13:00-14:00

SATELLITE LUNCH SYMPOSIUM

(unrestricted educational grant from Pfizer)

Managing haemophilia for life: transition in haemophilia

Chairs: Flora Peyvandi, Italy; Paula Bolton-Maggs, UK

- Emotional challenges of transition for people with life-long conditions. [Andrea Buzzi](#), Italy
- Transition in adolescence: treatment compliance and Quality of Life. [Paula Bolton-Maggs](#), UK
- Transition between products. [Charles Hay](#), UK

14:00-14:30 break

14:30-15:30

SATELLITE SYMPOSIUM

(unrestricted educational grant from Novo Nordisk)

Haemostasis in Novo Nordisk: present and future

Chairs: Flora Peyvandi, Italy; Elena Santagostino, Italy

- The best treatment for difficult-to-manage bleedings in patients with inhibitors. [Giovanni Di Minno](#), Italy
- R&D pipeline update. [Francesca Ferrante](#), Italy

14:00-14:30 break

15:30-16:00

SATELLITE LECTURE

(unrestricted educational grants from Swedish Orphan Biovitrum (SOBI) and Biogen Idec Hemophilia)

The Fc fusion technology for extended half-life of coagulation factor VIII and IX

Chairman: Stephen James, Sweden

- IgG Fc-fusions as therapeutic proteins – an overview. [Peter Lind](#), Sweden
- Biochemical and preclinical characterization of Fc fusions of factor VIII and factor IX with extended *in vivo* half-lives. [Robert Peters](#), USA

16:00-16:30 break

16:30-20:00

SCIENTIFIC SESSION 3

Chairs: Flora Peyvandi, Italy; Friz Scheifflinger, Austria

- Characterization of ADAMTS13 anti-ADAMTS13 antibody immune complexes in patients with acquired TTP. [Friedrich Scheifflinger](#), Austria
- The mechanism of VWF bond cleavage by ADAMTS13. [David Lane](#), UK
- Use of a murine model to assess biological function of ADAMTS13 and variants *in vivo*. [Xing Long Zheng](#), USA

18:00-18:30 break

- Differential proteolytic processing of oxidized VWF by ADAMTS-13 and leukocyte serine proteases. [Raimondo De Cristofaro](#), Italy
- Genetic cross-regulation of VWF and factor VIII antigen by SNPs. [Jing-Fei Dong](#), USA
- Genome-wide linkage and association studies of plasma VWF variation and other traits in healthy young adults. [Karl Desch](#), USA

Sunday, 22nd May

8:30-10:30

SCIENTIFIC SESSION 4

Chairs: Mary Scully, UK; Johanna Kremer-Hovinga, Switzerland

- Current treatments in TTP. [Mary Scully](#), UK
- New agents inhibiting the VWFGPIb alpha axis. [Samuel Machin](#), UK
- The link between complement and thrombosis in HUS. [Miriam Galbusera](#), Italy
- Atypical HUS as a paradigm to diagnosis and treatment of genetic diseases. [Elena Bresin](#), Italy

10:30-11:00 break

11:00-13:00

SCIENTIFIC SESSION 5

Chairs: Zaverio Ruggeri, USA; Ian Peake, UK

- Modelling thrombosis and haemostasis in the zebrafish. [Marguerite Neerman-Arbez](#), Switzerland
- Initiation and regulation of thrombogenesis at the vessel wall. [Zaverio Ruggeri](#), USA
- Role of VWF in ischemia/reperfusion injury. [Simone De Meyer](#), USA
- VWF regulates angiogenesis: cellular pathways and clinical implications. [Anna Randi](#), UK

13.00-14.00

SATELLITE LUNCH SYMPOSIUM

(unrestricted educational grant from Baxter)

From bench to bedside: development of rVWF and rADAMTS13

Chairs: Flora Peyvandi, Italy; Johanna Kremer Hovinga, Switzerland

- Implications of VWF and ADAMTS13 interaction. [Flora Peyvandi](#), Italy
- Development of recombinant ADAMTS13. [Friedrich Scheiflinger](#), Austria
- Development of rVWF: current status and future steps. [Tobias Suiter](#), Austria

14:00-14:30 break

14:30-15:00

SATELLITE LECTURE

(unrestricted educational grant from Bayer)

Chairman: Giovanni Di Minno, Italy

- Available tools to prolong half-life of factor VIII and VIIa. [David Lillicrap](#), Canada

15:00-15:30

SATELLITE LECTURE

(unrestricted educational grant from CSL Behring)

Chair: Elena Santagostino, Italy

- Innovations in recombinant coagulation factor technologies. [Gerhard Dickneite](#), Germany

15:30-19:30

SCIENTIFIC SESSION 6

Chairs: Augusto Federici, Italy, Giancarlo Castaman, Italy

- The historical aspects of VWD: diagnosis and classification. [Ian Peake](#), UK
- Fidelity of VWD diagnosis – from ristocetin to collagen. [Robert R. Montgomery](#), USA
- Type 1 VWD 10 years on. [Anne Goodeve](#), UK

17:00-17:30 break

- Reduced VWF survival in type 1 VWD. [Sandra Haberichter](#), USA
- Bleeding risk in VWD patients with increased VWF clearance. [Giancarlo Castaman](#), Italy
- Elevated VWF levels in VWD. [Reinhard Schneppenheim](#), Germany
- Prophylaxis in VWD - where do we stand today. [Erik Berntorp](#), Sweden

Monday, 23rd May

8:30-12:00

SCIENTIFIC SESSION 7

Chairs: Philip de Groot, the Netherlands; Peter Lenting, France

- Regulation of VWF activity. [Philip de Groot](#), the Netherlands
- Macrophage LRP modulates clearance of VWF. [Peter Lenting](#), France
- Storage and secretion of mutant VWF. [Jeroen Eikelboom](#), the Netherlands
- Functional analysis of VWF using knockout mice. [Cecile Denis](#), France

10:30-11:00 break

- Evaluating VWF interactions with GPIb alpha and ADAMTS13 in transgenic mouse models. [David Lillicrap](#), Canada
- The search for new GPIb-VWF inhibitors. [Hans Deckmyn](#), Belgium

12:00-13:00

POSTER DISCUSSION

Chairman: Pier Mannuccio Mannucci, Italy

- PO001 B CELL-ACTIVATING FACTOR (BAFF) IS ELEVATED IN ACUTE IDIOPATHIC THROMBOTIC THROMBOCYTOPENIC PURPURA (TTP)
Thomas M., Machin S.J., Mackie I., Scully M.A.
- PO002 ADAMTS13 ACTIVITY AND AUTOANTIBODIES CLASSES AND SUBCLASSES AS PROGNOSTIC PREDICTORS IN ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA (TTP)
Bettoni G., Palla R., Valsecchi C., Consonni D., Trisolini S., Carbone C., Peyvandi F.
- PO003 THROMBIN BINDING TO PLATELETS OF YOUNG PATIENTS WITH A HISTORY OF ACUTE MYOCARDIAL INFARCTION
Mendolicchio G., Bacci M., Zavalloni D., Rota L., Ruggeri Z.M.
- PO004 DEVELOPMENT OF A GAIN-OF-FUNCTION GPIBA BINDING ASSAY AS AN ALTERNATIVE TO THE RISTOCETIN COFACTOR ASSAY*
Pätzke J., Muth H., Winkens M., Althaus H., Budde U., Schneppenheim R.

- PO005 MULTIPLE DOMAINS MEDIATE LATERAL INCORPORATION OF VWF INTO ULVWF STRINGS ON THE SURFACE OF ENDOTHELIAL CELLS
Dragt B., Dienava I., De Laat B., Voorberg J.
- PO006 GPIB IN PLATELET-TYPE 2B VON WILLEBRAND DISEASE HAS A CONFORMATION THAT INCREASES ITS INTERACTION WITH THROMBIN: IMPLICATIONS FOR ENHANCED PLATELET ACTIVATION AND CONSUMPTION
Lancellotti S., Scaglione G., Arcovito A., Di Stasio E., Baronciani L., Peyvandi F., De Cristofaro R.
- PO007 DEFECTIVE COLLAGEN BINDING INTERACTION OF A NOVEL I1343V VON WILLEBRAND FACTOR GENE MUTATION
Castaman G., Giacomelli S.H., Jacobi P., Obser T., Budde U., Haberichter S. L., Schneppenheim R.
- PO008 CLINICAL EFFICACY AND SAFETY VERSUS BIOLOGICAL RESPONSE OF DESMOPRESSIN (DDAVP) IN INHERITED VON WILLEBRAND DISEASE TYPES 1 (VWD1) AND 2 (VWD2): RESULTS OF THE FIRST PROSPECTIVE AND INTERNATIONAL STUDY IN A COHORT OF 225 PATIENTS.
Federici A.B., Castaman G., Olivioecchio E., Blanchette V., Bonduel M., Mazzucconi M.G., Zieger B., D'Amico E., Lillicrap D., Lethagen S., Iorio A., Mannucci P.M.
- PO009 INCIDENCE AND DETERMINANT OF BLEEDING IN DIFFERENT TYPES OF VON WILLEBRAND DISEASE (VWD): RESULTS OF A PROSPECTIVE MULTICENTER COHORT STUDY ON 797 ITALIAN PATIENTS
Federici A.B., Bucciarelli P., Castaman G., Mazzucconi M.G., Morfini M., Rocino A., Schiavoni M., Rodeghiero F., Mannucci P.M.
- PO010 EXPRESSION STUDIES OF EIGHT MISSENSE MUTATIONS SPREAD THROUGH OUT THE VON WILLEBRAND FACTOR (VWF) IN TYPE 3 VON WILLEBRAND DISEASE (VWD) PATIENTS
Baronciani L., Solimando M., La Marca S., Cozzi G., Stufano F., Canciani M.T., Federici A.B., Peyvandi F.
- PO011 RUPTURE OF T CELL TOLERANCE TO SELF FVIII IN MILD HEMOPHILIA A.
Hermans C., Lavend'homme R., Saint-Remy J., Peerlinck K., Jacquemin M.
- PO012 TREATMENT OF BLEEDING EPISODES OCCURRING IN PATIENTS WITH ACQUIRED HAEMOPHILIA WITH FEIBA: THE EXPERIENCE IN ONE CENTRE (PADUA HAEMOPHILIA CENTRE)
Zanon E., Barbar S., Brandolin B., Saggiorato G., Spiezia L.

13.00-14.00

SATELLITE LUNCH SYMPOSIUM

(unrestricted educational grant from Grifols, Kedrion, LFB)

Immunogenicity of factor VIII products: clinical evidence

Chairman: Pier Mannuccio Mannucci, Italy

- Results of a new cohort study on the immunogenicity of different factor VIII products. [Elena Santagostino](#), Italy
- The SIPPET study: current status. [Pier Mannuccio Mannucci](#), Italy
- The RESIST studies. [Alessandro Gringeri](#), Italy

14:00-14:30 break

14:30-15:00

SATELLITE LECTURE

(unrestricted educational grant from Kedrion-LFB)

Chairman: A.B. Federici, Italy

- Wilfactin: more than ten years of successful clinical experience. [Jenny Goudemand](#), France

15:00-16:30

ORAL COMMUNICATIONS

Chairs: Pier Mannuccio Mannucci, Italy; Nicola Ciavarella, Italy

- OC001 INTERLEUKIN-28B (IL28B) POLYMORPHISM AS PREDICTOR OF SUSTAINED VIROLOGICAL RESPONSE AFTER ANTIVIRAL THERAPY FOR CHRONIC HEPATITIS C IN PATIENTS WITH HEMOPHILIA
Mancuso M., Aghemo A., Santagostino E., Rumi M., Bruno R., Colombo M., Mannucci P.M.
- OC002 UP-REGULATION OF PLATELET ACTIVATION IN HEMOPHILIA A
Van Bladel E.R., Roest M., De Groot P.G., Schutgens R.E.G.
- OC003 THE SHEAR STRESS-INDUCED TRANSCRIPTION FACTOR KLF2 AFFECTS DYNAMICS AND ANGIOPOIETIN-2 CONTENT OF WEIBEL-PALADE BODIES
Van Aghtmaal E., Bierings R., Fernandez-Borja M., Horrevoets A.J., Voorberg J.
- OC004 FREQUENCY OF HETEROZIGOUS GENE CONVERSION IN A COHORT OF 109 ITALIAN FAMILIES AFFECTED BY TYPE 2 VON WILLEBRAND DISEASE
La Marca S., Stufano F., Cozzi G., Garcia-Oya I., Canciani M.T., Baronciani L., Peyvandi F.
- OC005 DIFFERENT BLEEDING RISK IN TYPE 2 A AND 2 M VON WILLEBRAND DISEASE
Castaman G., Federici A., Baronciani L., Toseito A., Mannucci P.M., Rodeghiero F.
- OC006 CLINICAL, LABORATORY AND MOLECULAR MARKERS OF VON WILLEBRAND DISEASE TYPE 3 (VWD3): RESULTS FROM THE RETRO/PROSPECTIVE STUDIES ON 105/52 ITALIAN PATIENTS
Federici A.B., Bucciarelli P., Iorio A., Castaman G., Mazzucconi M.G., Morfini M., Rocino A., Schiavoni M., Olivovichio E., Mannucci P.M.
- OC007 THE PROGNOSTIC IMPLICATIONS OF ANTI-ADAMTS13 IGA, IGM AND IGG SUBCLASS DISTRIBUTION IN ACUTE IDIOPATHIC TTP
McDonald V., Mackie I.J., Scully M., Machin S.J.,
- OC008 CORRELATION BETWEEN SECRETION LEVELS OF ADAMTS13 MUTANTS AND FREQUENCY OF PLASMA TREATMENT IN CONGENITAL THROMBOTIC THROMBOCYTOPENIC PURPURA (TTP)
Rurali E., Banterla F., Bresin E., Galbusera M., Gastoldi S., Donadelli R., Noris M., Remuzzi G.
- OC009 DIFFERENT GLOMERULAR MRNA PROFILE OF COAGULATION-ASSOCIATED GENES BETWEEN DE NOVO AND RECURRENT RENAL THROMBOTIC MICROANGIOPATHY
Bockmeyer C.L., Agustian P.A., Lovric S., Schwarz A., Bröcker V., Becker J.U.

16:30-17:00 break

17:00-19:00

SCIENTIFIC SESSION 8

Chairs: W. Schramm, Germany; David Lillicrap, Canada

- Activation of the innate immune system is required for eliciting factor VIII inhibitor antibodies: towards an efficient prevention strategy. [Jean-Marie Saint-Remy](#), Belgium
- Induction of heme-oxygenase 1 reduces the development of factor VIII inhibitors. [Sebastien Lacroix-Desmazes](#), France
- Proteolytic antibodies in haemophilia: good news or bad news? [Srini V. Kaveri](#), France
- Issues of immunogenicity with current and future factor VIII products. [David Lillicrap](#), Canada

B

| | |
|--------------------|----|
| Baudo Francesco | 10 |
| Becker Ian Ulrich | 15 |
| Berntorp Erik | 12 |
| Bettoni Giuseppe | 13 |
| Bockmeyer Clemence | 15 |
| Bolton-Maggs Paula | 10 |
| Bresin Elena | 11 |
| Buzzi Andrea | 10 |

C

| | |
|--------------------|--------|
| Castaman Giancarlo | 12 |
| Ciavarella Nicola | 10, 14 |
| Coppo Paul | 10 |

D

| | |
|------------------------|----|
| De Cristofaro Raimondo | 11 |
| De Groot Philip G. | 13 |
| De Meyer Simone | 12 |
| Deckmyn Hans | 13 |
| Denis Cecile | 13 |
| Desch Karl | 11 |
| Di Minno Giovanni | 10 |
| Dickneite Gerhard | 12 |
| Dong Jing-Fei | 11 |
| Dragt Bleuwke | 13 |

E

| | |
|-----------------------|----|
| Eikenboom Jeroen C.J. | 13 |
|-----------------------|----|

F

| | |
|---------------------|------------|
| Federici Augusto B. | 12, 13, 14 |
| Ferrante Francesca | 10 |
| Fischer Kathleen | 10 |

G

| | |
|---------------------|----|
| Galbusera Miriam | 11 |
| Goodeve Anne | 12 |
| Goudemand Jenny | 14 |
| Gringeri Alessandro | 14 |

H

| | |
|--------------------|----|
| Hay Charles | 10 |
| Haberichter Sandra | 13 |

J

| | |
|----------------|----|
| Jacquemin Marc | 13 |
| James Stephen | 11 |

K

| | |
|---------------------------|--------|
| Kaveri Srin V | 15 |
| Kremer Hovinga Johanna A. | 10, 11 |

L

| | |
|----------------------------|------------|
| Lacroix-Desmazes Sebastien | 15 |
| La Marca Silvia | 14 |
| Lämmle Bernard | 10 |
| Lancellotti Luciano | 13 |
| Lane David | 11 |
| Lenting Peter | 13 |
| Lillicrap David | 12, 13, 15 |
| Lind Peter | 11 |

M

| | |
|-------------------------|--------|
| Machin Samuel | 10, 11 |
| Makris Mike | 10 |
| Mancuso Maria Elisa | 15 |
| Mannucci Pier Mannuccio | 10, 14 |
| Mendolicchio Loredana | 13 |
| McDonald Vickie | 15 |
| Montgomery Robert R. | 12 |

N

Neerman-Arbez Marguerite 12

P

Pätzke Jürgen 13

Peake Ian 12

Peters Robert 11

Peyvandi Flora 10, 11, 12

R

Randi Anna 12

Ruggeri Zaverio 12

Rurali Erica 15

S

Santagostino Elena 10, 14

Sørensen Benny 10

Stepanian Alain 10

Saint-Remy Jean-Marie 15

Scheiflinger Friedrich 11, 12

Schneppenheim Reinhard 12

Schramm Wolfgang 15

Scully Marie 11

Suiter Tobias 12

T

Thomas Mari 13

V

Vanhoorelbeke Karen 10

van Bladel Esther 14

van Agtmaal Ellen 14

Z

Zanon Ezio 13

Zheng Xing Long 11



7th Bari International Conference

Pugnochiuso, Vieste del Gargano, Foggia, Italy

22nd May 2011

h13:00-14:00

Satellite Symposium

From bench to bedside: development of rVWF and rADAMTS 13

Chairs

Johanna Kremer Hovinga, *Switzerland*; Flora Peyvandi, *Italy*

Agenda

Implications of von Willebrand factor and ADAMTS13 interaction

Flora Peyvandi, *Italy*

Development of recombinant ADAMTS13

Friedrich Scheifflinger, *Austria*

Development of rVWF: current status and future steps

Tobias Suiter, *Austria*