haematologica

the hematology journal

19th Congress of
the European Hematology Association
Milan, Italy, June 12 - 15, 2014

ABSTRACT BOOK

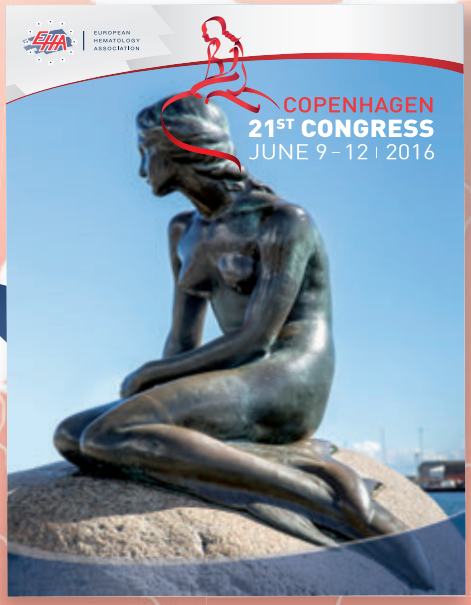
2014 | s1

ISSN 0390-6078
Journal of the European Hematology Association
Published by the Ferrata-Storti Foundation, Pavia, Italy
Volume 99, supplement no. 1, June 2014
www.haematologica.org

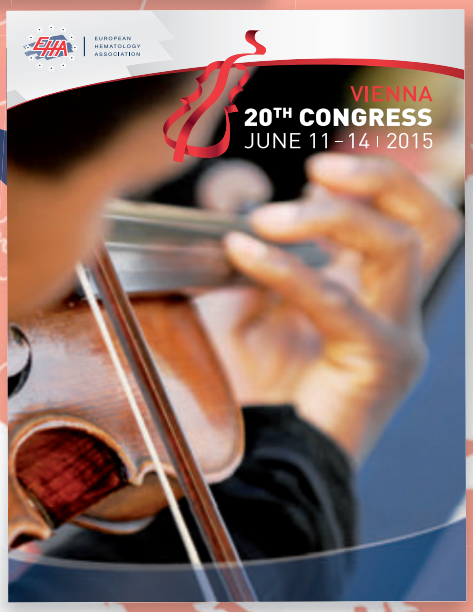
EUROPEAN
HEMATOLOGY
ASSOCIATION

FUTURE CONGRESSES

Copenhagen



Vienna



22nd Congress
Venue to be decided
June 22 - 25 2017

23rd Congress
Venue to be decided
June 14 - 17 2018

ehaweb.org

MILAN
19TH CONGRESS
JUNE 12 - 15 | 2014
European Hematology Association

19TH CONGRESS OF THE EUROPEAN HEMATOLOGY ASSOCIATION

MILAN, ITALY
JUNE 12-15, 2014

ABSTRACT BOOK

ISSN 0390-6078

The abstract book of the 19th Congress of the European Hematology Association is published as a supplement of Haematologica/the Hematology Journal in one volume per year.

All business correspondence and purchase and reprint requests should be addressed either to Haematologica Journal Office, via Giuseppe Belli 4, 27100 Pavia, Italy; phone: +39 0382 27129; fax: +39 0382 394705; e-mail: office@haematologica.org or to the European Hematology Association, Koninginnegracht 12b, 2514 AA The Hague, The Netherlands; phone: +31 (0)70 345 55 63; fax: + 31 (0)70 392 36 63; e-mail: info@ehaweb.org.

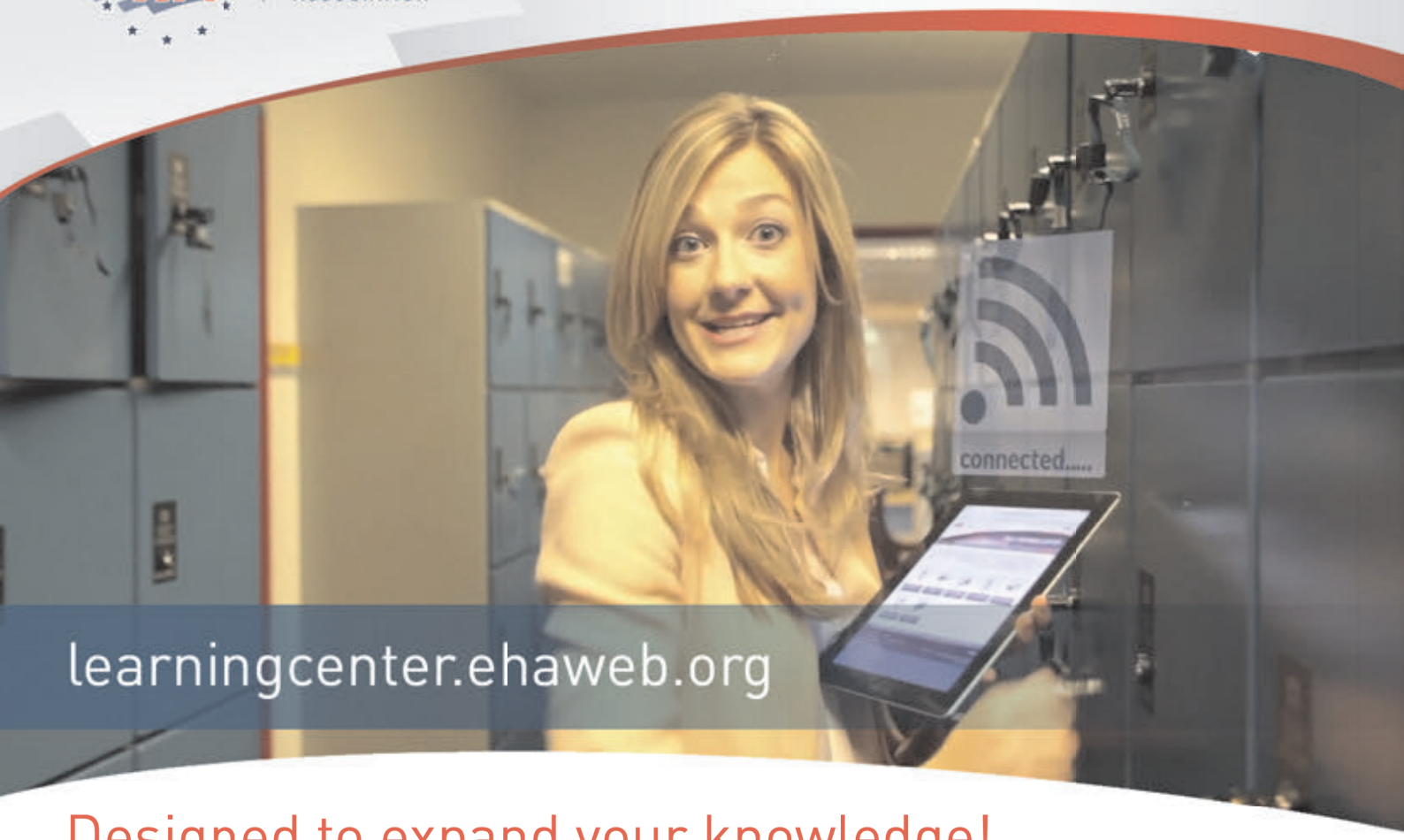
The Abstract book is available both at http://www.haematologica.org and http://www.ehaweb.org

Article Citations
Cite articles in this volume as follows:

TITLE. AUTHORS. JOURNAL YEAR; VOLUME(SUPPLEMENT NO):PAGE. Abstract n. XXX
Example: RITUXIMAB CONSOLIDATION AND MAINTENANCE THERAPY PROLONG RESPONSE DURATION IN B-CELL CHRONIC LYMPHOCYTIC LEUKEMIA
G. Del Poeta, M.I. Del Principe, A. Siniscalchi, L. Maurillo, F. Buccisano, A. Venditti, F. Luciano, P. Niscola, A. Zucchetto, V. Gattei, A.P. Perrotti, P. De Fabritiis, S. Amadori
Haematologica 2008; 93(s1):34. abstract n. 0085

haematologica
Journal of the European Hematology Association
Owned & published by the Ferrata Storti Foundation

EUROPEAN
HEMATOLOGY
ASSOCIATION



Designed to expand your knowledge!

The EHA Learning Center is accessible from your computer and mobile devices. All you need is an internet connection!

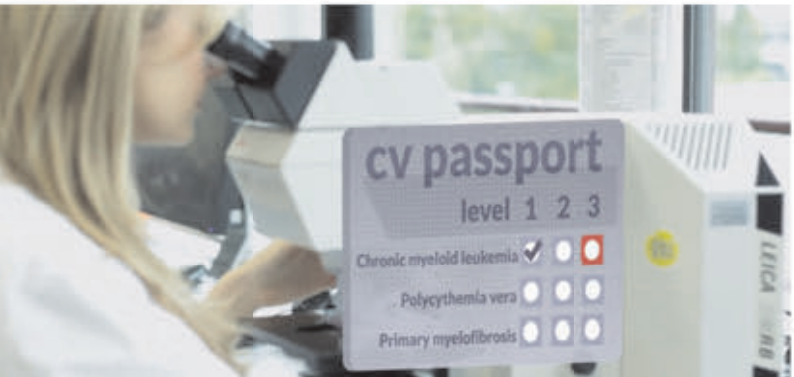
The Learning Center is fully linked to the EHA-CME System. Your credit points earned by successfully completing a CME activity, are directly uploaded into your personal CME account.

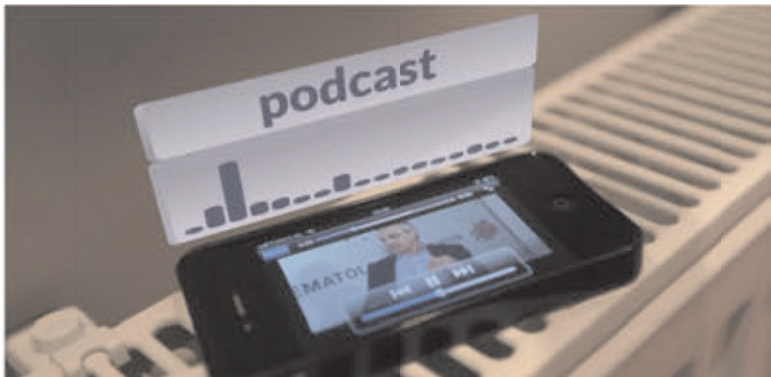
The EHA Learning Center offers a frame work of tools to deliver a centric learning experience by unifying theory, practice, technology and content.

Over the years, EHA has developed a number of online learning tools to facilitate easier access to knowledge for doctors. As a result, EHA's learning resources expanded into an extensive and user friendly platform.

It provides easy access to different ways of learning. All content on the EHA Learning Center is organized according to the European Hematology Curriculum.

See you soon at the EHA Learning Center!





EUROPEAN
HEMATOLOGY
ASSOCIATION

The European Hematology Association (EHA) promotes excellence in patient care, research and education in hematology.
By organizing an Annual Congress, providing an educational program based on the European Hematology Curriculum, publishing a journal, lobbying for hematology and promoting research, the Association supports hematologists in- and outside Europe to further improve hematology patient care.

Information about all the activities of the Association can be found on the website: ehaweb.org

EUROPEAN
HEMATOLOGY
ASSOCIATION

Raise the voice of European Hematology:

Become a member now!

Reduced fee of € 20 for Junior Members (up to 36 years of age)

Exclusive Membership Benefits are

- Free and unlimited access to EHA Learning Center
- Subscription to Haematologica/The Hematology Journal (impact factor 5.935 – monthly hard copy delivery)
- Eligibility to Career Development Program of EHA
- Eligibility to EHA Congress travel grants
- Discount on EHA Congress registration fee & all other EHA events
- Priority access to webcasts of EHA Congress
- Nomination and voting rights in EHA board member election
- Access to EHA membership directory
- Subscription to EHA newsletter (half-yearly hard copy delivery)
- Subscription to EHA e-bulletin

Apply online at ehaweb.org

MILAN
19TH CONGRESS
JUNE 12 - 15 | 2014
European Hematology Association

EHA Executive Board

C Chomienne, *France (President)*
J Sierra, *Spain (President Elect)*
U Jäger, *Austria (Past President)*
T Green, *United Kingdom (Secretary)*
P Sonneveld, *the Netherlands (Treasurer)*

EHA Councilors

A Brand, *the Netherlands*
D Bron, *Belgium*
J Cools, *Belgium*
A Engert, *Germany*
S Fröhling, *Germany*
D Grimwade, *United Kingdom*
L Malcovati, *Italy*
M Muckenthaler, *Germany*
G Ossenkoppele, *the Netherlands*
F Rodeghiero, *Italy*

Scientific Program Committee
19th Congress

P Sonneveld, *the Netherlands (Chair)*
E Angelucci, *Italy*
I Bodó, *Hungary*
A Brand, *the Netherlands*
R Delwel, *the Netherlands*
A Falanga, *Italy*
D Grimwade, *United Kingdom*
A Hochhaus, *Germany*
S Karlsson, *Sweden*
L Macintyre, *France*
M Mateos, *Spain*
C Montalban, *Spain*
J Sierra, *Spain*
G Socié, *France*
E Solary, *France*
K Stamatopoulos, *Greece*
S Stilgenbauer, *Germany*

Local Representative
Italian Society of Hematology

G Gaidano, *Italy*

Scientific Program Committee
Advisory Board 19th Congress

L Adès, *France*
A Balduzzi, *Italy*
M Beksac, *Turkey*
M de Bruijn, *United Kingdom*
H Dombret, *France*
S Eichinger, *Austria*
J Eikenboom, *the Netherlands*
B Falini, *Italy*
A Fielding, *United Kingdom*
S Fröhling, *Germany*
C Harrison, *United Kingdom*
M van den Heuvel, *the Netherlands*
A Husebekk, *Norway*
R Kralovics, *Austria/Czech Republic*
H Ludwig, *Austria*
C Moreno, *Spain*
F Morschhauser, *France*
E Papaemmanuil, *Greece*
JA Perez-Simon, *Spain*
J Porter, *United Kingdom*
A Reiter, *Germany*
G Saglio, *Italy*
A Sureda, *Spain*
A Taher, *Lebanon*
H de Thé, *France*
C-H Toh, *United Kingdom*
L Williamson, *United Kingdom*

EHA Executive Office

Koninginnegracht 12b
2514 AA The Hague
The Netherlands
Tel: +31 (0)70 345 55 63
Fax: + 31 (0)70 392 36 63
E-mail: info@ehaweb.org
Website: www.ehaweb.org

Abstract Reviewers

EHA would like to thank the following persons for their time and efforts reviewing abstracts for this Congress.

Adès L, France
Ageno W, Italy
Alberich Jorda M, Czech Republic
Almeida AM, Portugal
Amadori S, Italy
Ancliff P, United Kingdom
André M, Belgium
Angelucci E, Italy
Arat M, Turkey
Bahlis N, Canada
Balduzzi A, Italy
Barcellini W, Italy
Baron F, Belgium
Barrett J, USA
Beksac M, Turkey
Bellucci S, France
Bernd J, Austria
Bessler M, Philadelphia
Birgegard G, Sweden
Bjorkholm M, Sweden
Blaise D, France
Bodó I, Hungary
Boissel N, France
Bonnet D, France
Boultwood J, United Kingdom
Brand A, The Netherlands
Brice P, France
Brunet S, Spain
Bryder D, Sweden
Budde U, Germany
Bueren J, Spain
Bullinger L, Germany
Burnett A, United Kingdom
Buske C, Germany
Byrd J, USA
Casola S, Italy
Castagnetti F, Italy
Cazzaniga G, Italy
Cazzola M, Italy
Chase A, United Kingdom
Connors J, Canada
Constantinescu S, Belgium
Corey S, Chicago
Cosemans J, The Netherlands
Cosmi B, Italy
Czuczman M, USA
de la Rubia J, Spain
de Latour R, France
de Montalembert M, France
Delabesse E, France
Delforge M, Belgium
Delwel R, The Netherlands
Denis C, France
Döhner H, Germany
Dombret H, France
Donadieu J, France
Eichinger S, Austria
Eikenboom J, The Netherlands
Engelhardt M, Germany
Falanga A, Italy
Falini B, Italy
Falkenburg F, The Netherlands
Federico M, Italy
Fibbe W, The Netherlands
Fielding A, United Kingdom
Forni G, Italy
Fröhling S, Germany
Gaidano G, Italy
Gambacorti-Passerini C, Italy
Ganser A, Germany
Gardin C, France
Giagounidis A, Germany
Giebel S, Poland
Gisselbrecht C, France
Gonzalez-Barca E, Spain
Graux C, Belgium
Greinix H, Austria
Griesshammer M, Germany
Grimwade D, United Kingdom
Handgretinger R, Germany
Hantschel O, Switzerland
Hasle H, Denmark
Heidel F, Germany
Heidenreich O, United Kingdom
Hervig T, Norway
Heslop H, USA
Heuser M, Germany
Hochhaus A, Germany
Holler E, Germany
Holyoake T, United Kingdom
Hulin C, France
Hultcrantz M, Sweden
Husebekk A, Norway
Hutchings M, Denmark
Indrak K, Czech Republic
Ionnova T, Russia
Irving J, United Kingdom
Jäger U, Austria
Jelinek D, USA
Johnsen H, Denmark
Juliusson G, Sweden
Kappelmayer J, Hungary
Karlsson S, Sweden
Kattamis A, Greece
Kaufman J, USA
Kimby E, Sweden
Kjeldsen L, Denmark
Klein U, USA
Knop S, Germany
Korte W, Switzerland
Koschmieder S, Germany
Kosmider O, France
Kristinsson S, Sweden
Kumar S, USA
Kyrle P, Austria
Larocca A, Italy
Leleu X, France
Lenz G, Germany
Leprêtre S, France
Ljungman P, Sweden
Lübbert M, Germany
Luzzatto L, Italy
Machova K, Czech Republic
Macintyre E, France
Mahon FX, France
Malinge S, France
Martinez C, Spain
Martinez-Climent J, Spain
Mateos M, Spain
Meijer K, The Netherlands
Mendez-Ferrer S, Spain
Menendez P, Spain
Mills K, United Kingdom
Milsom M, Germany
Moccia A, Switzerland
Mohty M, France
Molina T, France
Montalban C, Spain
Montserrat E, Spain
Moreau P, France
Moreno C, Spain
Morotti A, Italy
Morschhauser F, France
Mufti G, United Kingdom
Munshi N, USA
Muszbek L, Hungary
Muus P, The Netherlands
Nagler A, Israel
Neri P, Canada
Nervi C, Italy
Neufeld E, USA
Nicolini F, France
Nissen C, Switzerland
Nomdedeu J, Spain
Oliva E, Italy
Ostermann H, Germany
Ott G, Germany
Papadaki H, Greece
Papaemmanuil E, Greece
Passamonti F, Italy
Perez-Simon JA, Spain
Petzer A, Austria
Piperno A, Italy
Puy H, France
Rahemtulla A, United Kingdom
Ramsay A, United Kingdom
Ravandi F, USA
Rebulla P, Italy
Reiter A, Germany
Remacha A, Spain
Risitano AM, Italy
Roberts D, United Kingdom
Robin C, The Netherlands
Rodeghiero F, Italy
Rousselot P, France
Rovó A, Switzerland
Rowntree C, United Kingdom
Saglio G, Italy
Sander B, Sweden
Santini V, Italy
Sanz G, Spain
Saussele S, Germany
Savage K, Canada
Scheding S, Sweden
Schlenk R, Germany
Schuringa JJ, The Netherlands
Seiffert M, Germany
Seifried E, Germany
Senis Y, United Kingdom
Shehata M, Austria
Sierra J, Spain
Sill H, Austria
Skokowa J, Germany
Socié G, France
Solal-Céligny P, France
Solary E, France
Sole F, Spain
Sonneveld P, The Netherlands
Stamatopoulos K, Greece
Stauder R, Austria
Steegmann JL, Spain
Stilgenbauer S, Germany
Strefford J, United Kingdom
Subklewe M, Germany
Sureda A, United Kingdom
Swinkels D, The Netherlands
Taher A, Lebanon
Thein SL, United Kingdom
Theurl I, Austria
Thielen N, The Netherlands
Toh CH, United Kingdom
Touw I, The Netherlands
Trka J, Czech Republic
Tuglular T, Turkey
Urbano-Ispizua A, Spain
Usmani S, USA
Uyl-de Groot C, The Netherlands
Van de Loosdrecht A, The Netherlands
van den Heuvel M, The Netherlands
van der Schoot E, The Netherlands
van Duin M, The Netherlands
Van Vlierberghe P, Belgium
Vanhoorelbeke K, Belgium
Verma A, USA
Vey N, France
Vigneri P, Italy
von Tresckow S, Germany
Vyas P, United Kingdom
Waage A, Norway
Walewski J, Poland
Watzke H, Austria
Weis J, Germany
Wendtner C, Germany
Williamson L, United Kingdom
Wilson W, USA
Zamagni E, Italy
Zeiser R, Germany

MILAN
19TH CONGRESS
JUNE 12 - 15 | 2014
European Hematology Association

Word of Welcome

On behalf of the EHA Board and the Scientific Program Committee we are pleased to present the Abstract Book of the 19th Congress of EHA.

The Scientific Program Committee has compiled an exciting and topical program of Simultaneous and Poster Sessions from over 2,300 submitted abstracts. Join our expert moderators for a walk along the (e)posters in your field of interest on Friday and Saturday and attend one of the 40 Simultaneous Sessions on Saturday and Sunday. The six Best Abstracts have been selected for presentation during the Presidential Symposium on Saturday afternoon.

In addition, please find at the end of the book also the late breaking abstracts that were submitted, reviewed and selected in April. The Scientific Program Committee is very excited about the quality of the studies represented in these abstracts and feels that they are a nice addition to the program. The eight selected oral presentations will be presented on Saturday in two Late Breaking Simultaneous Sessions. Furthermore, eleven posters are available for viewing in the Poster Area and will be presented on Saturday as well.

On behalf of the EHA Board, the committees and all the people involved in this years' EHA congress, we thank you for coming to Milan and wish you a good meeting.

Pieter Sonneveld
Chair Scientific Program Committee

haematologica
Journal of the European Hematology Association
Owned & published by the Ferrata Storti Foundation

Information for readers, authors and subscribers

Haematologica/The Hematology Journal (print edition, pISSN 0390-6078, eISSN 1592-8721) publishes peer-reviewed papers on all areas of experimental and clinical hematology. The journal is owned by a non-profit organization, the Ferrata Storti Foundation, and serves the scientific community strictly following the World Association of Medical Editors (WAME) recommendations on publication ethics policies for medical journals (www.wame.org/pubethicrecom. htm).

Haematologica/The Hematology Journal publishes editorials, perspectives, research papers, decision making & problem solving papers, review articles, brief reports and scientific letters. Manuscripts should be prepared according to the Uniform Requirements for Manuscripts Submitted to Biomedical Journals, prepared by the International Committee of Medical Journal Editors (ICMJE) and fully available online (http://www.icmje.org). Additional papers may be considered for the purely online journal (Haematologica/The Hematology Journal on Internet, eISSN 1592-8721). Because there are no space constraints online, Haematologica/The Hematology Journal will publish several items deemed by peer review to be scientifically sound and mainly useful as educational papers. These will include case reports, irreplaceable images, educational material from scientific meetings, meeting abstracts, and letters to the Editor.

Papers should be submitted online at http://www.haematologica.org/.

Conflict of interests. According to the International Committee of Medical Journal Editors (http://www.icmje.org/#conflicts), "Public trust in the peer review process and the credibility of published articles depend in part on how well conflict of interest is handled during writing, peer review, and editorial decision making". The ad hoc journal's policy is reported in detail online (http://www.haematologica.org/misc/about.dtl).

Galley Proofs and Reprints. Galley proofs should be corrected and returned by email, fax or express delivery within 48 hours. Minor corrections or reasonable additions are permitted; however, excessive alterations will require editorial re-evaluation and will be possibly charged to the authors. Papers accepted for publication will be printed without cost. The cost of printing color figures will be communicated upon request. Reprints may be ordered at cost by returning the appropriate form sent by the Publisher.

Transfer of Copyright and Permission to Reproduce Parts of Published Papers. Authors will grant copyright of their articles to the Ferrata Storti Foundation. No formal permission will be required to reproduce parts (tables or illustrations) of published papers, provided the source is quoted appropriately and reproduction has no commercial intent. Reproductions with commercial intent will require written permission and payment of royalties.

Detailed information about subscriptions is available online at URL http://www.haematologica.org. While access to the online journal is free, online access to some additional items available on http://www.haematologica.org may require either institutional or personal subscription.

Rates of the International edition for the year 2014 are as following:

	Institutional	*Personal*
Print edition (including full access to the online CME for personal subscribers)	*Euro 500*	*Euro 150*
Full access to the online CME only		*Euro 75*

To subscribe to the International edition, please visit our web site http://www.haematologica.org/misc/subscribe.dtl or contact: Haematologica Office, via Giuseppe Belli 4, 27100 Pavia, Italy (phone +39.0382.27129, fax +39.0382.394705, E-mail: office@haematologica.org).

Advertisements. Contact the Advertising Manager, Haematologica Office, via Giuseppe Belli 4, 27100 Pavia, Italy (phone +39.0382.27129, fax +39.0382.394705, e-mail: info@haematologica.org).

Disclaimer. Whilst every effort is made by the publishers and the editorial board to see that no inaccurate or misleading data, opinion or statement appears in this journal, they wish to make it clear that the data and opinions appearing in the articles or advertisements herein are the responsibility of the contributor or advisor concerned. Accordingly, the publisher, the editorial board and their respective employees, officers and agents accept no liability whatsoever for the consequences of any inaccurate or misleading data, opinion or statement. Whilst all due care is taken to ensure that drug doses and other quantities are presented accurately, readers are advised that new methods and techniques involving drug usage, and described within this journal, should only be followed in conjunction with the drug manufacturer's own published literature.

Direttore responsabile: Prof. Edoardo Ascari; Autorizzazione del Tribunale di Pavia n. 63 del 5 marzo 1955.
Printing: Tipografia PI-ME, via Vigentina 136, Pavia, Italy. Printed in June 2014.

Bleeding disorders

P615

CORRELATION BETWEEN OXIDATIVE STRESS AND BIOMARKERS OF JOINT DAMAGE IN PATIENTS WITH SEVERE HAEMOPHILIA TREATED BY DIFFERENT PROPHYLAXIS REGIMENS

I Djunic[1,*] V Dopsaj[2], P Miljic[1], N Suvajdzic-Vukovic[1], D Tomin[1], M Virijevic[1], A Novkovic[3], D Tomin[1], I Elezovic[1]
[1]Clinic for Hematology, Clinical Center of Serbia, [2]Institute for Medical Biochemistry, Pharmacy Faculty of Belgrade University, [3]Clinical Hospital Center "Zemun", Belgrade, Serbia

Background: Haemophilic arthropathy is the main cause of morbidity in individuals with severe haemophilia and prevention of joint disease is the primary goal of treatment. Recurrent bleeding into joints causes damage to all joint structures, but the precise mechanism by which this induces haemophilic arthropathy is still unclear. Recently, it was shown that one effect of blood in the joint is degradation of cartilage. Biomarkers of cartilage turnover can be measured in physiological fluids, such as plasma and urine.
Aims: The aims of this study were to detect correlation between a marker of oxidative stress -advanced oxidation protein products (AOPP) and serum/urine concentrations of biomarkers of joint cartilage degradation, as well as to estimate the influence of different prophylaxis regimens for severe haemophilia on this process.
Methods: The study included 20 adult patients with severe haemophilia, manifested by plasma factor (F)VIII/IX <1% of normal, without inhibitor. Five patients with haemophilia A received prophylaxis with FVIII concentrate in the standard dose of 20 IU/kg three times per week, while another five patients with haemophilia A were given an intermediate dose of FVIII concentrate as prophylaxis, 10-15 IU/kg thrice weekly. Seven patients with haemophilia A and three with haemophilia B, received FVIII/IX concentrate only on-demand. The following were measured: a) AOPP - a serum marker of oxidative stress and b) biomarkers of joint cartilage degradation - serum cartilage oligomeric matrix protein (COMP) and urinary C-terminal telopeptide of type II collagen (CTX-II). Blood and urine samples were collected initially, before the start of treatment (labelled AOPP-1, COMP-1 and CTX-II-1) and after 3 months follow-up (labelled AOPP-2, COMP-2 and CTX-II-2).
Results: The mean age of the patients was 32 years (range 19-55). In the group of patients given standard dose prophylaxis, the mean values of AOPP-2 (p=0.018), COMP-2 (p=0.043) and CTX-II-2 (p=0.014) were significantly lower than those for AOPP-1, COMP-1 and CTX-II-2. Likewise, the mean values for AOPP-2 (p=0.047) and CTX-II-2 (p=0.028) in the five patients receiving intermediate dose prophylaxis were also decreased when compared to initial values, but COPM level was not significantly changed. In patients treated on demand the mean values for AOPP, COMP and CTX-II did not alter significantly. The results showed marked positive correlations between AOPP and both COMP and CTX-II. Namely, lower values of AOPP were significantly associated with decreased levels of both biomarkers of cartilage degradation: COMP (p=0.008) and CTX-II (p=0.014).
Summary and Conclusions: The precise mechanism of joint disease in patients with severe haemophilia remains unknown but probably involves blood-induced increase of oxidative stress, which leads to higher joint cartilage turnover. The most important clinical strategy for management of these patients and prevention of severe arthropathy is treatment by continuous prophylaxis with intravenously applied FVIII/IX.

P616

BONE MINERAL DENSITY IN MEN AND CHILDREN WITH HAEMOPHILIA A AND B: A SYSTEMATIC REVIEW AND META-ANALYSIS

SA Paschou[1], P Anagnostis[2,*] SN Karras[1], C Annweiler[3], S Vakalopoulou[2], D Goulis[1], V Garipidou[2]
[1]Unit of Reproductive Endocrinology, First Department of Obstetrics and Gynecology, Medical School, [2]Haemophilia Center of Northern Greece, Aristotle University of Thessaloniki, Thessaloniki, [3]Department Geriatric Medicine, UPRES EA 4638, University Hospital, Angers, France; Robarts Research Institute, Department of Medical Biophysics, Schulich School of Medicine and Dentistry, the University of Western Ontario, London, Ontario, Canada, Angers, Greece

Background: Haemophilia is not considered among the classic causes of secondary osteoporosis.
Aims: The aim of this study was to systematically review the literature for case-control trials that have studied bone mass in males with haemophilia and to meta-analyze the best evidence available.
Methods: Electronic databases MEDLINE, EMBASE and CENTRAL were systematically searched for case-control trials that have studied bone mass in men or boys with haemophilia. Standardized mean difference (SMD) for bone mineral density (BMD) in the lumbar spine was the main study outcome and SMD in femoral neck and total hip BMD the secondary ones. Patient and control characteristics, such as age, body mass index (BMI), level of physical activity and blood-borne infections were recorded as possible predictors of the main outcome.
Results: Thirteen studies were included in the systematic review and 10 in the main outcome meta-analysis. Men with haemophilia demonstrated reduced lumbar spine [random effects SMD [95% confidence interval (CI)] -0.56 (-0.84; -0.28), between-study heterogeneity (I^2)=51%] and femoral neck BMD [random effects SMD (95% CI) -0.82 (-1.21; -0.44), I^2=63%] compared with controls, which indicated a large and clinically significant association. Similar results were obtained for children [random effects SMD (95% CI) -0.92 (-1.77; -0.07), I^2=92%]. No evidence of publication bias was detected. There was no evidence that age, BMI, level of physical activity or presence of blood-borne infections predicted lumbar spine BMD. Using the 'Common Language Effect Size' approach, the probability is about 72% that an individual without haemophilia would have higher lumbar spine BMD than an individual with haemophilia, if both individuals were chosen at random from a population.
Summary and Conclusions: This meta-analysis shows that men with haemophilia present a significant reduction in both lumbar spine and hip BMD, which appears to begin in childhood.

P617

CAN SECONDARY PROPHYLAXIS IN HAEMOPHILIA BE A CAUSE OF ISCHEMIA?

M Sonmez[1,*] H M Ozbas[1], A Mentese[2], D Us Altay[2]
[1]Haematology, [2]Biochemistry, Karadeniz Teknik University Medical Faculty, Trabzon, Turkey

Background: In the treatment of haemophilia, prophylactic factor is used to reduce bleeding frequency, to prevent complications and to improve quality of life. Life expectancy of patients with haemophilia improved with the advent of efficient management of bleeding attacks and widespread use of prophylactic treatment. The incidence of ischemic cardiovascular and cerebrovascular events is known to be lower compared to the general population due to the protective effect of hypocoagulable state associated with factor deficiency in haemophilia patients. The effect of factor administration during bleeding or of prophylactic treatment on ischemia development is not known. SCUBE 1, a new biochemical marker of protein structure, found in endothelial cells and platelets, is stored in α granules of platelets and released when platelets are stimulated. Levels of SCUBE 1 were found elevated in acute coronary syndrome and ischemic cerebrovascular events, making SCUBE 1 an early and potent indicator of ischemia.
Aims: The present study was designed to investigate the effect of factor treatment, SCUBE 1 levels.
Methods: Ten patients with haemophilia A and 3 with haemophilia B receiving prophylactic treatment were included into the study. Bloods samples were collected before and one hour after prophylactic administration. Additionally, during follow-ups, when bleeding episodes were observed blood samples were collected before and one hour after factor administration. In factor-treated cases, blood samples were drawn after administering the first 1000 units. Thirty healthy subjects were used as controls. Sera samples were stored at -80ºC and SCUBE 1 levels were studied using ELISA.
Results: Mean age for the patient and control groups were 35±10 and 34±7 years respectively. SCUBE 1 levels for groups were 235.4±20.3 ng/ml, 258.5±23.4 ng/ml and 235.9±16 in control, patients with bleeding before factor administration and patients before prophylaxis, respectively. SCUBE 1 levels were found to be 233.9±19.6 ng/ml and 280±29.7 ng/ml after factor administration in groups admitting for bleeding and prophylaxis, respectively. Statistically, patients admitting for bleeding had higher SCUBE 1 levels compared to controls, decreasing to that of control after factor administration ($p<0.01$). In prophylaxis group, SCUBE 1 levels of patients before factor administration were found to be comparable to that of controls, increased after factor administration ($p<0.0001$).
Summary and Conclusions: We conclude that, factor administration for prophylaxis may lead to ischemia, while administration in case of bleeding may have anti-ischemic behaviour.

P618

APPLICATION OF THE US NATIONAL INSTITUTE OF HEALTH (NIH) 2008 GUIDELINES FOR VON WILLEBRAND DISEASE IN A NATIONAL PAEDIATRIC COMPREHENSIVE CARE CENTRE.

I E Regan[1,*] A Burke[1], O M Cormack[1], C Fox[1], B Nolan[1]
[1]Coagulation/Haematology, Our Ladys Childrens Hospital, Crumlin, Dublin, Ireland

Background: Von Willebrand's Disease (VWD) is the most common inherited bleeding condition, affecting males and females in approximately equal proportions, and occurring in up to 1% of the world populations. Clinical and laboratory evaluation for VWD is relatively complex and there isno single laboratory test that can screen for the presence of VWD. The 2008 NHLBI/NIH guidelines on "The Diagnosis, Evaluation and Management of von Willebrand Disease" suggest an algorithmic approach to the clinical and laboratory diagnosis of VWD. These guidelines proposed more stringent diagnostic criteria to replace the 1995 and 2004 UKHCDO guidelines.