Program Outline

For an explanation of "Laptop session", Meet-the-Expert session" and "Clinical Challenges for Diagnosis" look at the end of the program.

Thursday 2 September 2010

16.30 Opening ceremony
Chairs: J.W.M. van der Meer, The Netherlands & A. Simon, The Netherlands

16.30 Opening
J.W.M. van der Meer, The Netherlands

16.45 Deficiency of Interleukin-1 receptor antagonist (DIRA)
I. Aksentijevich, USA

17.30 Art lecture
Gary Schwartz, Art historian

19.00 Walking dinner

Friday 3 September 2010

08.00 Breakfast Meet-the-Expert session 1
Clinical Phenotype of FMF
A. Livneh, Israel & M. Tunca, Turkey

Breakfast Meet-the-Expert session 2
Clinical Phenotype of HIDS
J.W.M. van der Meer, The Netherlands & J. Frenkel, The Netherlands, A. Prieur, France

Breakfast Meet-the-Expert session 3
Basic science Studying inflammasome
S. Masters, Ireland & V. Hornung, Germany

09.00 Plenary session: FMF + PAPA
Chairs: E. Ben-Chetrit, Israel & D. Kastner, USA

09.00 Environmental effects in FMF
S. Özen, Turkey

09.30 Regulation of MEFV gene expression
I. Touitou, France

10.00 The Effect of Colchicine on Pyrin-PSTPIP1 Interaction
Z.E. Taskiran, Turkey

10.15 Peripheral blood mononuclear cell (PBMC) cultures of M694V homozygous FMF patients reveal a predisposition to secrete high levels of IL-17 given a T cell activation signal.
Y. Shinar, Israel

10.30 Coffee break
11.00 **Plenary session: Oral presentations FMF + PAPA**  
*Chairs: E. Ben-Chetrit, Israel & D. Kastner, USA*

11.00 The effect of H. Pylori eradication in colchicine-resistant Familial Mediterranean Fever (FMF) Patients  
*G. Hakobyan, Armenia*

11.15 Validation of the newly developed pediatric criteria for the diagnosis of Familial Mediterranean fever in a large pediatric cohort of western European children with periodic fever  
*S. Federici, Italy*

11.30 Is familial mediterranean fever (FMF) associated with non-alcoholic fatty liver disease (NAFLD)?  
*M. Rozenbaum, Israel*

11.45 Analysis of MEFV exon methylation and expression patterns in familial mediterranean fever  
*E.T. Turanli, Turkey*

12.00 **Lunch**

12.00 **Laptop presentations**

13.00 **Plenary session: Clinical challenges for diagnosis: plenary discussion with expert panel**  
*Chair: J.P.H. Drenth, The Netherlands*

Speakers panel discussion:

- H. Yazici, Turkey, E. Ben-Chetrit, Israel, P. Hawkins, UK,  
- D. Kastner, USA, J.W.M. van der Meer, The Netherlands

14.30 **Plenary session: HIDS + TRAPS + PFAPA**  
*Chairs: J. Frenkel, The Netherlands & M. Gattorno, Italy*

14.30 IgD and immune surveillance  
*A. Cerutti, USA*

15.00 Adenotonsillectomy in PFAPA as first-line treatment: yes or no?  
*M. Hofer, Switzerland*

15.30 Analysis of an unfolded protein response and reactive oxygen species generation as a disease mechanism of TRAPS  
*J. Dickie, UK*

15.45 Reactive Oxygen Species Potentiate LPS-Mediated Inflammation in the TNF Receptor-Associated Periodic Syndrome (TRAPS)  
*A.C.B. Bulua, USA*

16.00 Discussion

16.15 **Tea break**

16.45 **Plenary session: Oral presentations HIDS + TRAPS + PFAPA**  
*Chairs: J. Frenkel, The Netherlands & M. Gattorno, Italy*

16.45 TRAPS associated mutations affect a multitude of intracellular pathways beyond the canonical TNFR1 pathways through alterations in both protein abundance and signalling response dynamics.  
*P.J. Tighe, UK*

17.00 PFAPA Syndrome – a systems biology approach reveals activation of the innate and adaptive immunity with
responsiveness to IL-1β blockade
S. Stojanov, Germany
17.15 TLR2/TLR4-dependent aberrant IL-1 signaling in hyperimmunoglobulinemia D and periodic fever syndrome (HIDS)
M. Stoffels, The Netherlands
17.30 Role for Rac1 signaling in the development of autoinflammatory disease
R. Van der Burgh, The Netherlands

Saturday 4 September 2010

08.00 Breakfast Meet-the-Expert session 4
All about genetics
I. Touitou, France & J.P.H. Drenth, The Netherlands
Breakfast Meet-the-Expert session 5
Clinical Phenotype of CAPS
R. Goldbach-Mansky, USA & H. Hoffman, USA
Breakfast Meet-the-Expert session 6
Basic Science Studying Cytokines
M. Netea, The Netherlands & C. Dinarello, USA

09.00 Plenary session: Regulation of innate immunity
Chairs: C. Dinarello, USA & M. Netea, The Netherlands
09.00 Dissecting IL-1 and caspase activation
T. Kanneganti, USA
09.30 Inflammasome activation by dsDNA
V. Hornung, Germany
10.00 Inflammasome activation
G. Guarda, Switzerland
10.30 Inflammasome-dependent and -independent activation of IL-1beta
M. Netea, The Netherlands

11.00 Coffee break

11.30 Plenary session: Oral presentations Regulation of innate immunity
Chairs: C. Dinarello, USA & M. Netea, The Netherlands
11.30 Temperature sensitive TRP channels in human peripheral blood mononuclear cells.
M. Stoffels, The Netherlands
11.45 Endothelial-derived secondary necrotic apoptotic bodies release biologically active IL-1alpha
P. Kaplanski, France
12.00 Pattern recognition receptors in normal and inflamed human epidermis: upregulation of dectin-1 in psoriasis
H.D. de Koning, The Netherlands
12.15 Do NALP3 play a role in cytoplasmatic HIV recognition and dendritic cells activation, linking innate to specific immunity in HIV infection?
A. Pontillo, Italy
12.30  Lunch

12.30  Laptop presentations

13.30  Workshop 1
Basic science issues in the lab: how to measure inflammasome activation / IL-1 processing?
Chair: H. Waterham, The Netherlands
Speakers panel discussion: C. Dinarello, USA, M. Netea, The Netherlands, A. Rubartelli, Italy

Workshop 2
International Patient Registries
Chair: J. Frenkel, The Netherlands
Speakers panel discussion: M. Gattorno (Eurofever), Italy, H. Lachmann (Eurotraps), UK, M. Hofer (PFAPA), Switzerland, H. de Koning (Schnitzler), The Netherlands

14.30  Plenary session: Autoinflammation versus autoimmunity
Chair: J.P.H. Drenth, The Netherlands
Panel members: D. Kastner, D. McGonagle, J. Cohen Tervaert & J. van der Meer

16.00  Tea break

16.30  Plenary session: CAPS / DIRA / NALP12
Chairs: R. Goldbach-Mansky, USA & H. Hoffman, USA

16.30  In vivo IL-1 beta regulation in CAPS
H. Lachmann, UK

17.00  Long-term followup of anakinra in CAPS
R. Goldbach-Mansky, USA

17.30  Rilonacept in CAPS
H. Hoffman, USA

18.00  Coffee break

18.15  Plenary session: Oral presentations CAPS / DIRA / NALP12
Chairs: R. Goldbach-Mansky, USA & H. Hoffman, USA

18.15  Functional consequences of a germline mutation in the leucin rich repeat domain of NLRP3 identified in an atypical autoinflammatory disorder
I. Jéru, France

18.30  Hearing loss in Muckle-Wells-Syndrome – effects of IL-1 inhibition
J.B. Kuemmerle-Deschner, Germany

18.45  Initial characterization of a mouse model of neonatal onset multisystem inflammatory disease
D. Brydges, USA

19.00  A redox signature shared by NLRP3-mutated and NLRP12-mutated monocytes causes accelerated secretion of IL-1beta
A. Rubartelli, Italy

20.00  Congress dinner
Sunday 5 September 2010

08.30  **Workshop 3**  
Animal Models for diseases  
*Chair: H. Waterham, The Netherlands*  
Mouse model FMF  
J. Chae, USA  
Mouse model CAPS  
S. Brydges, USA  
Mouse model HIDS  
*H. Waterham, The Netherlands & M. Gibson, USA*  
Mouse model CRMO  
*P. Ferguson, USA*  
Mouse model TRAPS  
*R. Siegel, USA*

**Workshop 4**  
Genetics: quality control and genetic tests  
*Chair: I. Touitou, France*  
*Panel members:*  
I. Aksentijevich, USA  
Y. Shinar, Israel  
J. Arostegui, Spain

10.00  **Coffee break**

10.30  **Plenary session: Other systemic autoinflammatory diseases**  
*Chairs: J.W.M. van der Meer, The Netherlands & H. Yazici, Turkey*  
10.30  Treatment of Behcet’s disease  
*H. Yazici, Turkey*  
11.00  **The EMBO lecture:** Heart failure as a reactive autoinflammatory disease  
*C. Dinarello, USA*  
11.30  Classification of Immunological Disease-Focus on MHC Class 1 intermediate disorders  
*D. McGonagle, UK*

12.00  **Plenary session: Oral presentations Other systemic autoinflammatory diseases**  
*Chairs: J.W.M. van der Meer, The Netherlands & H. Yazici, Turkey*  
12.00  IL-18 Binding protein (il-18bp) dramatically improves liver histological lesions in an animal model of macrophage activation syndrome (mas).  
*P. Kaplanski, France*  
12.12  PED-BD: An international cohort study on pediatric Behçet’s disease: One-year data of 110 patients  
*I. Koné-Paut, France*  
12.24  Granulomas in NOD2-related Pediatric Granulomatous Arthritis and Crohn’s Disease: an Immunohistochemical Study
C.E.I. Janssen
12.36  Mapping dynamic protein interactions of Receptor Interacting Protein 2 (RIP2) and ProCaspase-1 by co-immunoprecipitation and life-cell imaging
S.R. Hofmann, Germany

12.48  Clinical phenotype and CARD15 gene mutation with Blau Syndrome in Chinese children and their parents
C.F. Li, China

13.00  Lunch

13.00  Laptop presentations

14.30  Plenary session: Developments in treatment and diagnostics
Chairs: H. Lachmann, UK & S. Özen, Turkey
14.30  S100A12 protein
H. Wittkowski, Germany
15.00  Alpha1-antitrypsin
C. Dinarello, USA
15.30  Developments in IL-1 inhibition
H. Lachmann, UK

16.00  Tea break

16.30  Plenary session: Oral presentations Developments in treatment and diagnostics
Chairs: H. Lachmann, UK & S. Özen, Turkey
16.30  Colchicine does not decrease the need for immunosuppressive use at long term in behçet’s syndrome (bs)
G. Hatemi, Turkey
16.42  Safe, Rapid-onset, and Sustained Biological Activity of IL-1ß Regulating Antibody XOMA 052 in Resistant Uveitis of Behçet’s Disease: Preliminary Results of a Pilot Trial
A. Gul, Turkey
16.54  The role of montelukast in the management of hyperimmunoglobulinemia D with periodic fever syndrome(HIDS)
P. Goldsmith, USA
17.06  Experience with anakinra in 120 patients with systemic autoinflammatory diseases
A. Melo Gomes, Portugal
17.18  On demand rather than continuous anakinra treatment is effective in mevalonate kinase deficiency
E.J. Bodar, The Netherlands

17.30  Plenary session: Oral presentations New Genes and Late Break-through
Chairs: A. Simon, The Netherlands & D. Kastner, USA
17.30  Functional human protein analysis in the post-genome era learning to new autoinflammatory syndrome (Nakajo-Nishimura syndrome)
H. Ida, Japan
17.45  Identification of the mutation predisposing to a periodic fever syndrome in Chinese Shar-Pei dogs
M. Olsson, Sweden
18.00  A novel autoinflammatory syndrome with lymphedema (aisle)
Monday 6 September 2010

08.00  Breakfast Meet-the-Expert session 7
       Clinical Phenotype of TRAPS
       R. Powell, UK & P. Hawkins, UK

08.00  Breakfast Meet-the-Expert session 8
       Treatment of FMF
       H. Ozdogan, Turkey & E. Pras, Israel

08.00  Breakfast Meet-the-Expert session 9
       Clinical Phenotype of PFAPA
       M. Hofer, Switzerland & S. Stojanov, USA

09.00  Plenary session: Amyloidosis, other long-term complications and prognosis
       Chairs: B. Hazenberg, The Netherlands & P. Hawkins, UK
09.00  New insights development of AA amyloidosis
       J. van der Hilst, The Netherlands
09.30  Pharmacological depletion of SAP
       P. Hawkins, UK
10.00  Atherosclerosis and the inflammasome in DM type 2
       S. Masters, Ireland

10.30  Coffee break

11.00  Plenary session: Oral presentations Amyloidosis, other long-term complications and prognosis
       Chairs: B. Hazenberg, The Netherlands & P. Hawkins, UK
11.00  Evaluation of autonomic nervous system dysfunction in FMF patients, with and without amyloidosis
       U. Nussinovitch, Israel
11.15  Causes and predictors of death and prognosis in familial mediterranean fever in the colchicine era
       G. Can, Turkey
11.30  AA Amyloidosis Complicating the Inherited Periodic Fever Syndromes
       T. Lane, UK

11.45  Closing plenary session
       Chairs: J. Frenkel, The Netherlands & J.W.M. van der Meer, The Netherlands
11.45  3 best laptop presentations
12.15  Closing session
       D. Kastner, USA

12.45  Closing
Clinical Challenges for Diagnosis
Establishing the diagnosis of a specific autoinflammatory disease can be very challenging. A few of such diagnostic challenges will be presented to teams of renowned clinicians that will try to solve them during a public discussion on Saturday September 4. You are kindly invited to provide us with complicated cases, where a diagnosis has been reached, or as yet unsolved cases.
Deadline for submission: first of August 2010.
Please contact us (e.erren@congresscare.com) if you have a suitable case in mind. It is possible to stay anonymous if you prefer, since the aim is purely to examine the diagnostic process in a potential case of autoinflammatory disease.
Friday September 3
13.00 – 14.30

Laptop sessions
The usual poster sessions will be substituted by laptop sessions. Instead of bringing a printed-out poster, each laptop presenter will bring his/her own laptop. In the designated area, each presenter will be assigned a standing-table where he or she can talk their audience through their work, using approximately 5 powerpoint slides. The audience will have the opportunity to walk freely between the several laptop presenters. We hope this setup will encourage face-to-face interaction and discussion. For participants who have no laptop, we will provide a small number of laptops on location - please let us know in advance if you need this.
Friday September 3
12.00 – 13.00
Saturday September 4
12.30 – 13.30
Sunday September 5
13.00 – 14.30

Meet-the-expert-sessions
These are informal and interactive sessions. The invited experts may prepare a short presentation if they want to, but the main goal of these sessions is to get a chance to ask questions, discuss dilemmas and exchange ideas. On the on-line registration form, we ask you to choose the sessions you would like to participate in.
Friday September 3
08.00 – 09.00 Meet-the-Expert session 1, 2 & 3
Saturday September 4
08.00 – 09.00 Meet-the-Expert session 4, 5 & 6
Monday September 6
08.00 – 09.00 Meet-the-Expert session 7, 8 & 9