

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. Aksentjevich, 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-1 β 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-1 β
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-1 α
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 Riloncept 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 leucine 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

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 live-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-1 β 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)

- G. Hatemi, Turkey*
- 18.15 Genome-wide Association Study Identifies Common Variants of the IL10 and IL23R Genes that Contribute to Behçet Disease Susceptibility
F. Remmers, USA

Monday 6 September 2010

- 08.00 **Breakfast Meet-the-Expert session 7**
Clinical Phenotype of TRAPS
R. Powell, UK & P. Hawkins, UK
- Breakfast Meet-the-Expert session 8**
Treatment of FMF
H. Ozdogan, Turkey & E. Pras, Israel
- 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**

A. Simon, The Netherlands

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