



POSTER PRESENTATION

Open Access

Evidence based recommendations for diagnosis and treatment of cryopyrin-associated periodic syndromes (CAPS)

Nienke Ter Haar^{1*}, Marlen Oswald², Luca Cantarini³, Marco Gattorno⁴, Michael Hofer⁵, Isabelle Kone-Paut⁶, Jordi Anton-Lopez⁷, Karyl Barron⁸, Paul Brogan⁹, Joost Frenkel¹⁰, Caroline Galeotti¹¹, Gilles Grateau¹², Veronique Hentgen¹³, Tilmann Kallinich¹⁴, Helen Lachmann¹⁵, Huri Ozdogan¹⁶, Seza Ozen¹⁷, Anna Simon¹⁸, Yosef Uziel¹⁹, Carine Wouters²⁰, Brian Feldman²¹, Bas Vastert²², Nico Wulffraat²², Susanne Benseler²³, Jasmin Kümmerle-Deschner²

From 21st European Pediatric Rheumatology (PReS) Congress
Belgrade, Serbia. 17-21 September 2014

Introduction

Cryopyrin-associated periodic syndromes (CAPS) is a group of rare monogenetic autoinflammatory disorders. Evidence-based guidelines are lacking and management is mostly based on physician's experience. Consequently, treatment regimens differ throughout Europe. In 2012, a European initiative called SHARE (*Single Hub and Access point for pediatric Rheumatology in Europe*) was launched to optimize and disseminate diagnostic and management regimens in Europe for children and young adults with rheumatic diseases.

Objectives

One of the aims of SHARE was to provide evidence based recommendations for management (treatment and monitoring) of CAPS.

Methods

Evidence based recommendations were developed using the European League Against Rheumatism (EULAR) standard operating procedure. An expert committee was instituted, consisting of pediatric and adult rheumatologists. The expert committee defined search terms for the systematic literature review. Two independent experts scored articles for validity and level of evidence. Recommendations derived from the literature were evaluated by an online survey. Those with less than 80% agreement on the

online survey or with relevant comments of the experts were reformulated. Subsequently, all recommendations were discussed at a consensus meeting using Nominal Group Technique. Recommendations were accepted if more than 80% agreement was reached.

Results

The literature search yielded 1698 articles, of which 25 papers on treatment were considered relevant and therefore scored for validity and level of evidence. Seventeen were scored valid and used in the formulation of the recommendations. Fifteen recommendations were suggested in the online survey and discussed during the consensus meeting. Six general recommendations on management, five for monitoring and four for treatment were accepted with more than 80% agreement. Topics covered are the following: the multidisciplinary team, treatment goals, adjunctive therapies, psychosocial support and vaccinations [general recommendations], monitoring frequency, minimal assessments in all CAPS patients and monitoring of severe phenotypes [monitoring] and IL-1 blockade, NSAIDs and/or glucocorticoids during attacks and DMARDS/biologicals other than IL-1 blockade [treatment].

Conclusion

The SHARE initiative provides recommendations for the management of CAPS and thereby facilitates improvement and uniformity of care throughout Europe.

¹Laboratory for Translational Immunology, University Medical Center Utrecht, Utrecht, Netherlands

Full list of author information is available at the end of the article

Disclosure of interest

N. Ter Haar Grant / Research Support from: SHARE is funded by the European Commission (project N° 20111202), M. Oswald: None declared., L. Cantarini Grant / Research Support from: Novartis, SOBI, Consultant for: Novartis, SOBI, M. Gattorno Grant / Research Support from: Novartis, Speaker Bureau of: SOBI, M. Hofer: None declared., I. Kone-Paut Grant / Research Support from: Chugai, Novartis, SOBI, Consultant for: Abbvie, Chugai, Novartis, Pfizer, SOBI, Speaker Bureau of: Novartis, Pfizer, J. Anton-Lopez Grant / Research Support from: Abbvie, Novartis, Pfizer, Consultant for: Novartis, Speaker Bureau of: Abbvie, Novartis, Pfizer, Roche, SOBI, K. Barron: None declared., P. Brogan Grant / Research Support from: Novartis, Roche, Consultant for: Novartis, J. Frenkel Consultant for: Novartis, Speaker Bureau of: SOBI, C. Galeotti Grant / Research Support from: Novartis, G. Gâteaux Consultant for: Novartis, V. Hentgen Consultant for: Novartis, T. Kallinich Grant / Research Support from: Novartis, Speaker Bureau of: Novartis, SOBI, H. Lachmann: None declared., H. Ozdogan: None declared., S. Ozen Consultant for: Novartis, Speaker Bureau of: Biovitrium, A. Simon Consultant for: Novartis, SOBI, Xoma, Y. Uziel Grant / Research Support from: Novartis, Consultant for: Novartis, Speaker Bureau of: Abbvie, Neopharm, Novartis, Roche, C. Wouters: None declared., B. Feldman: None declared., B. Vastert Consultant for: Novartis, N. Wulffraat Grant / Research Support from: Abbvie, GSK, Roche, Consultant for: Genzyme, Novartis, Pfizer, Roche, S. Benseler: None declared., J. Kümmerle-Deschner Grant / Research Support from: Novartis, Speaker Bureau of: SOBI.

Authors' details

¹Laboratory for Translational Immunology, University Medical Center Utrecht, Utrecht, Netherlands. ²Klinik für Kinder- und Jugendmedizin, Abteilung für pädiatrische Rheumatologie, Autoinflammation Reference Center Tübingen, Universitätsklinikum Tübingen, Tübingen, Germany. ³Policlinico Le Scotte, University of Siena, Siena, Italy. ⁴G. Gaslini Institute, Genova, Italy. ⁵Department of Pediatrics, University of Lausanne and, University of Geneva, Switzerland. ⁶Department of Pediatric Rheumatology, Reference Centre for Autoinflammatory Disorders CEREMAI, Bicêtre Hospital, University of Paris SUD, Paris, France. ⁷Hospital Sant Joan de Déu, Universitat de Barcelona, Barcelona, Spain. ⁸NIH, Bethesda, USA. ⁹Department of Rheumatology, UCL Institute of Child Health, London, UK. ¹⁰Department of Pediatrics, University Medical Center Utrecht, Utrecht, Netherlands. ¹¹Reference Centre for Autoinflammatory Disorders CEREMAI, Bicêtre Hospital, University of Paris SUD, Paris, France. ¹²Centre national de référence des amyloses d'origine inflammatoire et de la fièvre, Hôpital Tenon, AP-HP, université Pierre-et-Marie-Curie, Paris, France. ¹³Centre Hospitalier de Versailles, Le Chesnay Cedex, France. ¹⁴Charité University Medicine, Berlin, Germany. ¹⁵National Amyloidosis Centre, University College London Medical School, London, UK. ¹⁶Cerrahpasa İc Hastalıkları Kliniği, Istanbul Turkey. ¹⁷Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey. ¹⁸Department of Medicine, Radboudumc, Nijmegen, Netherlands. ¹⁹Department of Pediatrics, Sapir Medical Center, Kfar Saba, Tel-Aviv University, Sackler School of Medicine, Tel-Aviv, Israel. ²⁰University Hospital Leuven, Leuven, Belgium. ²¹Division of Rheumatology, The Hospital for Sick Children, Toronto, Canada. ²²Department of Pediatric Immunology, University Medical Center Utrecht, Utrecht, Netherlands. ²³The Hospital for Sick Children, Toronto, Canada.

Published: 17 September 2014

doi:10.1186/1546-0096-12-S1-P78

Cite this article as: Haar et al.: Evidence based recommendations for diagnosis and treatment of cryopyrin-associated periodic syndromes (CAPS). *Pediatric Rheumatology* 2014 **12**(Suppl 1):P78.

Submit your next manuscript to BioMed Central and take full advantage of:

- Convenient online submission
- Thorough peer review
- No space constraints or color figure charges
- Immediate publication on acceptance
- Inclusion in PubMed, CAS, Scopus and Google Scholar
- Research which is freely available for redistribution

Submit your manuscript at
www.biomedcentral.com/submit

