



POSTER PRESENTATION

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

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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.

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Disclosure of interest

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