



POSTER PRESENTATION

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# Therapy of autoinflammatory diseases: results from an international registry

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From 18th Pediatric Rheumatology European Society (PReS) Congress  
Bruges, Belgium. 14-18 September 2011

## Background

The evidence for therapy in autoinflammatory diseases is limited. There are few randomized controlled trials or disease registries.

## Aim

To evaluate the response to treatment of autoinflammatory diseases based on data of an international registry.

## Methods

A web-based registry collecting baseline and clinical information of autoinflammatory diseases was located at the member area of the PRINTO website ([www.printo.it](http://www.printo.it)). Participating hospitals included pediatric rheumatology centers of the PRINTO network and adult centers with a specific interest in autoinflammatory diseases. Data was collected on Blau's syndrome, Behçet's disease, CAPS, CRMO, DIRA, FMF, MKD, NLRP12-mediated periodic fever, PAPA, PFAPA TRAPS and undefined periodic fevers. In total, 704 patients were included in this retrospective study on therapy.

## Results

NSAIDs and steroids were beneficial in most diseases. Anakinra induced a complete response in 70% of 40 treated CAPS patients, 82% of 28 TRAPS patients and in both DIRA patients. Furthermore, it was beneficial in 80% of 10 MKD patients. Etanercept was completely effective in all 6 Behçet patients, 34% of 32 TRAPS patients and 13% of 8 MKD patients and partially effective in 53% of TRAPS and 38% of MKD patients. Colchicine was beneficial in approximately 95% of 131 FMF

and 18 Behçet patients, although the complete response rate was just 56% and 22%, respectively. For PFAPA syndrome, corticosteroids aborted the attacks in 78% of 113 patients.

## Conclusion

The results were compared to the literature. These combined findings could serve as a base for therapeutic guidelines and identify candidate drugs for future therapeutic trials.

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Published: 14 September 2011

doi:10.1186/1546-0096-9-S1-P18

Cite this article as: ter Haar et al.: Therapy of autoinflammatory diseases: results from an international registry. *Pediatric Rheumatology* 2011 **9** (Suppl 1):P18.

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