







## Ottava Giornata della Ricerca della Svizzera Italiana Venerdì 9 marzo 2018

## Modulo per la sottomissione abstract di ricerca CLINICA

Titolo (massimo 15 parole)

THROUGH NEW CLASSIFICATION CRITERIA FOR INHERITED PERIODIC FEVERS AND PFAPA

Autori (cognome e iniziali, es: Grassi L.)

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**Testo** (massimo **250 parole**, preferibilmente in italiano (accettato anche in inglese), suddiviso in Introduzione, *Metodi, Risultati, Conclusioni* e *Finanziamento* 

Objective. To develop classification criteria for hereditary periodic fevers (HPF) and PFAPA.

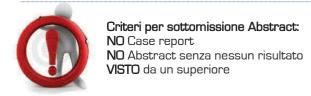
Methods. A multistep process, based on a combination of expert consensus and analysis of real patient data, was conducted. A panel of experts (clinicians and geneticists) was asked to classify 360 patient profiles as having or not one of the four HRF (FMF, TRAPS, MKD, CAPS) or PFAPA, based on clinical, genetic and laboratory features collected in the Eurofever registry. The 360 patients comprised 60 patients for each disease and 60 patients with undifferentiated autoinflammatory disease with recurrent fever. Next, the sensitivity, specificity, positive predictive value, negative predictive value and area under the curve of candidate criteria, derived from a previous Delphi survey and univariate/multivariate analysis, were assessed. The best performing criteria for each disease were selected and presented and discussed in a consensus conference.

Results. Experts achieved consensus on the classification of 281/360 (87%) patient's profiles. 193 candidate criteria with high specificity and sensitivity were selected and evaluated in the consensus conference. During the conference, experts arrived to a consensus on 2 sets of criteria for each HRP, one including genetic and clinical variables, the other with clinical variables only. One clinical criteria for PFAPA was also selected.

Conclusion. We have developed a set of classification criteria for HPF and PFAPA with high specificity and sensitivity. Prospective validation is required to confirm the high accuracy of the criteria.

**Visto superiore** (prego indicare Nome e Cognome del superiore)

Prof. Giacomo Simonetti



**Invio Abstract**