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Perspective validation of the eurofever classification criteria for monogenic periodic fevers

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Introduction

We recently proposed a set of provisional, evidence-based, clinical criteria for the classification of children and adults patients affected by monogenic periodic fevers. These criteria, built and validated in a cohort of 1215 patients with periodic fever enrolled in the Eurofever Registry from November 2009 to February 2013, displayed a high sensitivity and specificity.

Objectives

To prospectively validate the provisional Eurofever clinical classification criteria in an independent set of patients with periodic fevers enrolled in the registry after March 2013

Methods

The provisional Eurofever classification criteria (best accuracy cut-off) were applied to patients with periodic fevers enrolled in the registry after March 2013. The following disease were analyzed: FMF, MKD, TRAPS, CAPS, PFAPA and undefined periodic fevers. Patients were divided in three subgroups: i) group A: inherited periodic fevers with genetic confirmation, ii) group B: inherited periodic fevers with non-confirmatory genetic test, iii) group C: negative controls. Statistical analysis compared the sensitivity and specificity of best accuracy cut-off in patients belonging to group A and C.

Results

A total of 245 patients were evaluated. Among them 80 were classified in group A (35 FMF, 28 MKD, 9 TRAPS, 8 CAPS), 94 in group B (56 FMF, 6 MKD, 9 TRAPS, 23 CAPS) and 71 in group C (59 PFAPA and 12 undefined periodic fever). The classification criteria tested on patients belonging to Group A and C displayed an overall good sensitivity and specificity (respectively 85% and 92% for FMF, 92% and 89% for MKD, 100% and 96% for CAPS and 100% and 93% for TRAPS). (see table 1). The performance of the classification criteria in patients with non-confirmatory genetic test are reported in Table 1 below.

Table 1

	Sensitivity (Best Accuracy cut-off)				Specificity (Best Accuracy cut-off)			
	FMF	**MKD**	**CAPS**	**TRAPS**	**FMF**	**MKD**	**CAPS**	**TRAPS**
Genetically confirmed (Group A)	85%	92%	100%	100%	92%	89%	96%	93%
Not genetically confirmed (Group B)	58%	67%	45%	25%	94%	79%	100%	82%

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Conclusion

Perspective validation of Eurofever classification criteria showed their good accuracy in patients with confirmatory genetic test. Despite a lower sensitivity, the criteria displayed an high specificity in the classification of patients with non-confirmatory molecular test. In order to evaluate the actual accuracy of the classification criteria in this latter subgroup, an expert validation is currently ongoing.

Disclosure of interest

None declared.

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