Validation of the PEDiatic Behçet Disease Criteria (PEDBD): a consensus-based approach

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Introduction: In children only one set of classification exists, known as PEDBD criteria (2015), created by an international Expert consensus. It is still preliminary and requires improvement in both specificity and sensitivity.

Objectives: To perform an external validation of the PEDBD criteria

Methods and results:

Random selection of 210 pediatric patients: BD, PFAPA, FMF, MKD, TRAPS, SURF and UND

Exclusion of the patients included in the 2015 PEDB criteria

1° Round: clinical & serological data shown

consensus reached in 45/210

2° Round: genetic data shown

consensus reached in 58/163

Total consensus on 103/210 (49%)

3° Round: Experts’ votes & comments shown

EXPERT CONSENSUS AS GOLD STANDARD DEFINITION

application of the PEDBD, ISG and ICBD criteria to the cohort of classified BD and to the other diseases in order to evaluate their performance on BD and confounding diseases

Conclusions:

- Pediatric Behçet disease classification showed a wide range of opinions among Experts.
- The disease heterogeneity is a real challenge in practice but the drafting of a more robust classification may allow to identify homogeneous groups of patients for research purpose and possibly discriminate a larger number of differential diagnoses.

BD, Behçet disease; FMF familial mediterranean fever; MKD mevalonate kinase deficiency; PFAPA periodic fever, aphthosis, pharyngitis, adenitis; TRAPS TNF receptor associated periodic syndrome, SURF sdr of undifferentiated recurrent fever; UND undefined inflammatory diseases