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PReS-FINAL-2319: PED-BD cohort 2013: expert consensus classification gives higher sensitivity than the international study group criteria to define Behcet's disease in children

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Introduction

BD is rarely encountered in children where the disease is very difficult to recognize. The outcome of patients with few symptoms is currently unknown

Objectives

To define the outcome of paediatric patients with at least two symptoms of BD, and to obtain an appropriate definition of BD in patients <16y.

Methods

An international expert committee has defined the criteria of inclusion. New patients or patients followed for a maximum of 3 years, who presented at least 2 symptoms of BD (among a list), and gave their informed consent were included, reviewed yearly.

Results

228 patients were included since 2008, (SR: 1), from 22 centres of 13 countries, median age of 12.5y. Median age at first symptom was 7.2y. Family history of BD was present in 22% and consanguinity in 4.5%. Median disease duration at inclusion was 4.7y and from the first symptom to last visit was 7.5y. Inclusion criteria plus oral aphtosis (mandatory) were (%): genital aphtosis 50, necrotic folliculitis 31, uveitis 28, familial history 22, pathergy positive 19, erythema nodosum 15, vascular 10 and retinal vasculitis 7.

Mean number of symptoms: 1 plus family history 41%, 2 (33%), more than 3 (26%). Patient had a median of 1.3 follow-up visit (0-4). 220 patients had a first visit, 138 patients had a 1-y visit (mean BD duration: 5.8y). 81 patients had 2-y (6.4y), 44 a 3-y (7.3y) and 18 a 4-y visit (7.5y). The symptoms along the study were (%): dermatological 67, genital aphtosis 52, articular 48, fever 47, gastrointestinal 39, ocular 36, neurological 35, pathergy 17, vascular 12, urological 2. HLAB51 was present in 47%. Male patients had significantly more ocular and vascular signs, female had more genital aphtosis. Between 1st-4th visit: 57% had no new symptom, 24% had 1, 11% had 2 and 10% had more than 3. The expert committee has examined 199 files at a median disease duration of 6,1y, and classified 121 patients as definite, 18 as probable and 3 as not BD. 57 charts were reviewed but did not reached consensus. 46 files have been reviewed more than once.. Among our patients classified as definite: 121/142 (85%); 79/121 (65%) fulfilled the ISG International criteria. International criteria and expert classification showed significant differences. Although good concordance (Kappa c = 0.72). Having 2 or more symptoms was significantly associated with classification as definite BD (p = 0.0005).

Conclusion

The expert committee has classified the majority of patients in the BD group although they did not fulfil the international BD classification criteria (for adults).

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Disclosure of interest

None declared.

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