Basic and translational science in paediatric rheumatology.

CHILDHOOD ABDOMINAL/PELVIC PAIN AS A PREDICTOR OF RECURRENT MUSCULOSKELETAL PAIN IN ADOLESCENT BOYS AND GIRLS: A PROSPECTIVE POPULATION-BASED STUDY

Keywords: Epidemiology, Pain

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LEUCOCYTE ABNORMALITIES AND CYTOKINE LEVELS IN SYNOVIAL FLUID OF JUVENILE IDIOPATHIC ARTHRITIS PATIENTS: A PRELIMINARY STUDY

Keywords: Cell biology, Cytokines and chemokines, Uveitis

Disclosure of Interests: None Declared.

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Paediatric rheumatology

POS0740
VALIDATION OF THE PEDIATRIC BEHÇET DISEASE CRITERIA (PEDBD): A REAL LIFE CONSENSUS-BASED APPROACH

Keywords: Vasculitis, Behcet’s disease

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Background: Behçet’s syndrome (BS) is an autoinflammatory disease characterized by a variable vessel vasculitis. In children, BS may start early in life, mimicking other autoinflammatory diseases and making the diagnosis challenging. In the past, several criteria have been created for adult BS classification. In 2015, the first set of BS paediatric classification criteria, the PEDBD, was proposed by an international Expert consensus [1].

Objectives: to perform an external validation of the PEDBD criteria in a cohort of internationally validated paediatric BS patients.

Methods: 210 patients (70BS, 40 PFAPA, 35 FMF, 26 MDK, 22 TRAPS, 17 UND/ SURF) were randomly selected from the Eurofever Registry (patients excluded if > 16 years and if included in first PEDBD study). A set of 11 Experts blinded to original diagnosis, were chosen to evaluate the patients: in the 1st round, clinical and serological data were evaluated; in the 2nd round genetic diseases and Immunodeficiencies, Genoa, Italy

Results: At the end of the 3rd round, a consensus on the initial diagnosis was reached in 112/210 patients (53.3%), with an additional consensus on a different final diagnosis in 27 patients. The BS patients with an agreement (24) were classified as confirmed-BS, and those with an agreement of 60-70% (10) as probable-BS. In confirmed-BS patients, oral ulcers were present in all the patients, genital ulcers in 77%, skin manifestations in 50%, a positive pathergy test in 39%, anterior and posterior uveitis in 29 and 27%, retinal vasculitis and papillary oedema in 8% of patients, with a resulting impaired vision in 17%. Venous thrombosis was present in 2 patients (8%). The patients with ocular and vascular involvement were all males. Cranic nerve palsy (17%) was the most frequent neurologic symptom. Abdominal pain was present in 33%, diarrhoea and gastrointestinal bleeding in 13% and anal/perianal ulcers in 8%. 29% presented arthralgia, and 13% arthritis. Fever was present in 50% of patients. HLA-B51 was positive in 69% of patients. When comparing these patients with the confounding diseases, an older age at disease onset, the presence of oral and genital ulcers, skin papulo-pustular lesions, a positive pathergy test and posterior uveitis were BS distinctive elements. The ISG, ICBD and PEDBD criteria were applied to confirmed and probable-BS and to the confounding disease controls, resulting in the following test characteristics (Table 1).

Table 1.

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>Accuracy</th>
</tr>
</thead>
<tbody>
<tr>
<td>ISG</td>
<td>0.50</td>
<td>1.0</td>
<td>0.90</td>
</tr>
<tr>
<td>ICBD</td>
<td>0.79</td>
<td>0.98</td>
<td>0.95</td>
</tr>
<tr>
<td>PEDBD</td>
<td>0.58</td>
<td>0.99</td>
<td>0.91</td>
</tr>
</tbody>
</table>

Conclusion: the PEDBD criteria were extremely specific but had a lesser sensitivity than ICBD which had a better accuracy. One limitation is that specific monogenic BS mimics were not included as disease controls, thus the true accuracy of all these criteria may be lower in practice. The complexity of childhood BS suggests that genotyping (incorporating autoinflammatory diseases, BS mimics, and HLA-type) combined with clinical features are likely to yield the most accurate classification criteria, which would require further validation in a larger cohort.


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POS0741
CLINICALLY UNEXPECTED JOINT INFLAMMATION DETECTED BY WBMRI IS MORE FREQUENT IN JIA PATIENTS THAN CONTROLS

Keywords: Immunological arthritis, Outcome measures, Imaging

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Background: Whole-body MRI (WBMRI) enables a comprehensive assessment of joint inflammation in children and young people (CYP) with juvenile idiopathic arthritis (JIA). The clinical relevance of WBMRI-detected inflammation in joints requires further assessment.

Objectives: To evaluate the frequency of WBMRI-detected subclinical joint inflammation in CYP with JIA and without inflammatory arthritis. To explore the relationship between WBMRI-detected and clinically detected joint inflammation in patients with JIA.

Methods: CYP aged 14-24 with JIA (any subtype) or non-inflammatory joint pain controls) were prospectively recruited in a cross-sectional study. All participants underwent a Dixon-based WBMRI scan after being clinically assessed. Based on clinical findings, the CYP with JIA were divided into the active (≥1 active joint or sacroiliitis) and inactive group (no active joints or sacroiliitis). Three musculoskeletal radiologists blindly and independently reviewed the post-contrast images for joint inflammation, and this was considered present if detected by ≥2 radiologists. Periarticular joint inflammation was defined as above-normal intensity synovial enhancement with ≥1 additional feature (synovial hypointensity, subarticular oedema, joint effusion or periarticular soft tissue oedema). The frequency of subclinical joint inflammation (WBMRI-detected inflammation in ≥1 clinically inactive joint per participant) was compared between the control and JIA groups, and between the JIA subgroups using unpaired proportions.

Results: 47 CYP with JIA and 13 controls were included. The median age of participants was 17 years (IQR 16 to 20) with 62% and 85% female in JIA and controls respectively. Twenty-seven (57%) participants with JIA were treated with biologic disease-modifying anti-rheumatic drugs (DMARDs) and 29 (62%) with conventional synthetic DMARDs. The frequency of WBMRI-detected joint inflammation was higher in active than inactive JIA patients (Table 1). A higher percentage of patients with JIA were detected to have subclinical inflammation by WBMRI (49%, 23/47) compared to controls (15%, 2/13), difference of 34% (95% CI: 9, 58%). One joint with inflammation was detected by WBMRI in both controls, respectively. Fifty-six per cent (14/25) of JIA patients labelled active clinically had WBMRI-detected subclinical inflammation, compared to 41% (9/22) of clinically inactive JIA patients (Table 1). The proportion of patients classified as active or inactive by both clinical and WBMRI assessment varied between JIA subtypes (Figure 1).

Conclusion: This study demonstrated that WBMRI-detected subclinical joint inflammation is a common finding in JIA, but rare in controls. Inflammation is more common in patients with clinically active compared with inactive disease.