PEDIATRIC RHEUMATOLOGY

ORAL PRESENTATION Open Access

Patients with early-onset systemic juvenile idiopathic arthritis show more inflammation and worse outcome

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From 18th Pediatric Rheumatology European Society (PReS) Congress
Bruges, Belgium. 14-18 September 2011

Background
Systemic Juvenile Idiopathic Arthritis (SJIA) is heterogeneous in severity, course and outcome. Predictive factors for a poor outcome include persistent systemic features (fever, thrombocytosis) and younger age at onset.

Aim
To describe and analyze the disease features in patients with SJIA with very early onset and to compare them with those of patients with later onset.

Methods
Retrospective analysis of clinical data. Early-onset (EO) was defined as the start of SJIA prior to age 18 months. Variables included: demographic and clinical features at onset and outcome variables during disease course (pattern of course, presence of clinical joint damage [using the Juvenile Arthritis Damage Index or JADI], radiographic joint damage [erosions], destructive hip disease, disability [CHAQ > 0.5], growth retardation, development of macrophage activation syndrome [MAS], need for biologic agents, and death). Patients with EO were compared with patients with disease onset at age ≥ 18 months. Chi square and Mann-Whitney tests were used for comparisons.

Results
192 patients (115 girls, 23 EO) followed between 1995 and 2010 were included. Age at onset was 12 (2-17) months in patients with EO, 72 (18-191) months in patients with non-EO. Delay in diagnosis (2 months) and duration of follow-up (10 vs 8 years) were similar in both groups. Eight patients (1 EO, 7 non-EO) died during the observation period. Patients with EO showed more frequently serositis (p=0.0003), hepatomegaly (p=0.01), splenomegaly (p=0.03), lower number of active joints (p=0.01), Hgb (p=0.02), and higher plt (p=0.04) at onset; more frequently MAS (p=0.0001), therapy with biologics (p=0.01), destructive hip disease (p=0.04), radiographic damage (p=0.01), growth retardation (p=0.05), disability (p=0.01), and higher JADI score (p=0.003).

Conclusions
Patients with SJIA starting before age 18 months show more systemic inflammatory features, and a poorer outcome than children with later disease onset. They may represent a distinct subset. This observation should prompt rheumatologists to initiate early aggressive therapy and close follow-up in this age group.

Published: 14 September 2011

doi:10.1186/1546-0096-9-S1-O18
Cite this article as: Russo and Katsicas: Patients with early-onset systemic juvenile idiopathic arthritis show more inflammation and worse outcome. *Pediatric Rheumatology* 2011 **9**(Suppl 1):O18.

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