Abstract

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**Objective. To develop a composite disease activity score for systemic JIA (sJIA) and to provide preliminary evidence of its validity.**

**Methods. The systemic Juvenile Arthritis Disease Activity Score (sJADAS) was constructed by adding to the four**

**items of the original JADAS a fifth item that aimed to quantify the activity of systemic features. Validation analyses**

**were conducted on patients with definite or probable/possible sJIA enrolled at first visit or at the time of a flare,**

**who had active systemic manifestations, which should include fever. Patients were reassessed 2 weeks to 3 months**

**after baseline. Three versions were examined, including ESR, CRP or no acute-phase reactant.**

**Results. A total of 163 patients were included at 30 centres in 10 countries. The sJADAS was found to be feasible and to possess face and content validity, good construct validity, satisfactory internal consistency (Cronbach’s alpha 0.64–0.65), fair ability to discriminate between patients with different disease activity states and between those whose parents were satisfied or not satisfied with illness outcome (P < 0.0001 for both), and strong responsiveness to change over time (standardized response mean 2.04–2.58). Overall, these properties were found to be better than those of the original JADAS and of DAS for RA and of Puchot score for adult-onset Still’s disease.**

**Conclusion. The sJADAS showed good measurement properties and is therefore a valid instrument for the as asessment of disease activity in children with sJIA. The performance of the new tool should be further examined in other patient cohorts that are evaluated prospectively.**