What is low-dose corticosteroid therapy in juvenile idiopathic arthritis?
A worldwide, questionnaire-based survey*

**Resultate:** Von 99 zurückgeschickten Fragebogen waren 92 auswertbar. Die im Rahmen einer Langzeittherapie noch als niedrig angesehene Kortikosteroiddosis betrug im Mittel 0,26 ± 0,14 mg Prednisolon/kgKG/d (min-max = 0,04–0,50 mg, n = 92). Dabei waren die Mittelwerte aus Nordeuropa (0,29 ± 0,12, n = 9), Westeuropa (0,42 ± 0,14, n = 7), Südeuropa (0,30 ± 0,14, n = 9), Osteuropa (0,25 ± 0,14, n = 6) und Nordamerika (0,33 ± 0,17, n = 16) höher als aus Mitteleuropa (0,19 ± 0,09, n = 43).

**Schlussfolgerung:** Die Vorstellungen der Kinderhautologen, was unter einer niedrigdosierten Kortikosteroid-Langzeittherapie zu verstehen sei, unterscheiden sich im internationalen Vergleich um den Faktor zehn. Die Ursache dieser bemerkenswerten Differenz und die Auswirkungen der unterschiedlichen Behandlung auf den Langzeitverlauf sollten untersucht werden. Eine international akzeptierte Definition einer „low-dose, long-term corticosteroid therapy“ sollte entwickelt und dann in prospektiven Studien überprüft werden.

**Schlüsselwörter** Juvenile idiopathische Arthritis – Therapie – Kortikosteroide – Dosierung

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**Dedication:**
Dedicated to Frau Prof. Dr. med. Elisabeth Stoeber on the occasion of her 90th birthday.

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* Instead of the old terms “juvenile rheumatoid arthritis” or “juvenile chronic arthritis”, the new ILAR-WHO nomenclature “juvenile idiopathic arthritis” is used throughout the paper.

**Summary Objective:** To determine pediatric rheumatologists’ personal definitions of systemic low-dose, long-term (>4 weeks) corticosteroid therapy of juvenile idiopathic arthritis (JIA).

**Methods:** Pediatric rheumatologists from America, the Near East (Israel), Australia and Europe were asked for their personal definition of a low-dose long-term corticosteroid therapy of JIA with the aid of a standardized questionnaire.

**Results:** Of 99 questionnaires returned, 92 were evaluable. The dosage still considered low turned out to be 0,26 ± 0,14 mg prednisolone/kgBW/day (min-max = 0,04–0,50 mg, n = 92). Higher dosages were indicated from Northern Europe (0,29 ± 0,12, n = 9), Western Europe (0,42 ± 0,14, n = 7), Southern Europe (0,30 ± 0,14, n = 9), Eastern Europe (0,25 ± 0,14, n = 6) and North America (0,33 ± 0,17, n = 16) than from Central Europe (0,19 ± 0,09, n = 43).

**Conclusion:** Pediatric rheumatologists’ personal definitions of low-dose, long-term corticosteroid therapy vary within a wide range. The reason for these differences and the impact on patients should be investigated. In addition, a generally accepted definition for low-dose, long-term corticosteroid therapy should be developed and subsequently examined in studies.

**Key words** Juvenile idiopathic arthritis – therapy – corticosteroids – dosage
Introduction

For the systemic, long-term treatment of juvenile idiopathic arthritis (JIA), corticosteroids are only being used in a rather restricted manner because of their various adverse effects, including in part irreversible growth suppression (5, 6, 8, 10). While higher dosages are mandatory in critical situations, like severe myocarditis, only a low-dose regimen is acceptable for long-term anti-arthritic therapy.

Kirwan et al. have demonstrated that low-dose corticosteroid therapy can slow down the radiologic progression of rheumatoid arthritis (3, 4). However, there is still no proof that this holds true for the corticosteroid therapy of JIA as well. Moreover, there is no generally accepted definition to date for “low-dose” during a systemic, long-term corticosteroid therapy of JIA.

This paper presents the results of a worldwide questionnaire survey. The personal definitions of 92 pediatric rheumatologists of “low-dose corticosteroid therapy” for JIA differ by a factor of ten.

Methods

By means of a questionnaire, pediatric rheumatologists were asked, “Which doses do you consider low, average or high during systemic, long-term treatment (> 4 weeks) of juvenile chronic/rheumatoid arthritis (mg prednisolone/kg body weight/24h)?” The questionnaire was initiated on the occasion of a lecture on the corticosteroid therapy of JIA given at the “5th European Conference on Pediatric Rheumatology”, held in Garmisch-Partenkirchen on October 15–19, 1997 (6). The questionnaire contained the following three additional questions; the results thereof are not reported here: 1) Indications? 2) Which preparations do you prefer? 3) Do you use a pulse therapy? If yes, what is your method?

The participants were additionally asked whether they would agree to the anonymous publication of data and whether their names may or should be listed in a publication. Only questionnaires with the agreement of the participant were used for the evaluation presented here.

The questionnaires were sent to pediatric rheumatologists in Europe, America, Australia and Israel. Selected were pediatric rheumatologists who were known to be leading experts of their particular country from publications and from international meetings (ACR, EULAR, European conferences on pediatric rheumatology). In Germany, the questionnaire was sent to all members of the “Arbeitsgemeinschaft für Kinder- und Jugendrheumatologie” who are known to treat children with JIA on a regular basis.

Results

From the 125 questionnaires, 99 (79 %) were returned and 92 (93 %) of these were analyzable. In seven out of the 99 questionnaires returned, no prednisolone dosages per kg body weight were listed, but only daily total dosages with no relation to body weight. The participants were free to name a single figure or a range as being “low dose”. Seventy-two participants listed a single figure while 20 provided a range. In case a range was given, the upper figure was used for the calculation of the mean value, standard deviation, minimal and maximal values, median and spread (Table 1).

The mean for “low dose” of the 92 questionnaires was 0.26 ± 0.14 mg of prednisolone/kgBW/day (Table 1). The data vary

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a Austria, Germany, Switzerland  
b Denmark, Finland, Norway, Sweden  
c France, Netherlands, UK  
d Bulgaria, Italy, Greece, Portugal, Spain, Turkey  
e The Czech Republic, Hungary, Lithuania, Poland, Russia  
f Canada, USA  
g Brazil, Chile

Table 1 Results of a questionnaire-based survey with the question “Which doses do you consider low during systemic, long-term treatment of juvenile chronic/rheumatoid arthritis (mg prednisolone/kg body weight/24h)?”. (n number of participants; x mean value; s standard deviation; Min minimal value; Max maximal value).