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The Relationship Between Impairments in Joint Function and Disabilities in Independent Function in Children with Systemic Juvenile Idiopathic Arthritis

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ABSTRACT.

. Objective. Knowledge about the impact of joint impairment on functional ability is needed in planning care and setting treatment goals in children with juvenile idiopathic arthritis (JIA). We investigated the relationship between joint impairments and upper and lower limb function.

Methods. Twenty-one children with systemic JIA with an average age of 9.2 years and a mean disease duration of 4.8 years participated in this study. Joint impairments were assessed by the following variables: joint counts on swollen (JCS) and tender (JCT) joints and the loss of joint motion as determined by the Joint Alignment and Motion scale (JAM). Functional performance and functional ability were determined by the Juvenile Arthritis Functional Assessment Scale (JAFAS) and Childhood Health Assessment Questionnaire (CHAQ), respectively. The relationship between impairments and functional disabilities was studied at the level of (1) the complete instruments, (2) upper and lower limb function separately, and (3) the individual joints and items.

Results. Regarding complete instruments, the Spearman rank correlation between functional disabilities and loss of joint motion was moderate to good (JAM/CHAQ $r_s = 0.66$, JAM/JAFAS $r_s = 0.77$).

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A fair correlation was found between functional disabilities and the joint count on swollen joints (JCS/CHAQ $r_s = 0.45$, JCS/JAFAS $r_s = 0.52$), but no significant relationship was found with the number of tender joints (JCT/CHAQ $r_s = 0.02$, $p > 0.05$, and JCT/JAFAS $r_s = 0.14$, $p > 0.05$). At the extremity level (upper and lower limb function), the relationship between functional disabilities and the loss of joint motion appeared to be stronger in the leg than in the arm. At the level of the individual joints and questionnaire items, loss of joint motion in hip or shoulder joint appeared to be the most important factor in predicting limitation in leg or arm function.

Conclusion. Our study shows that with respect to joint impairments, loss of joint motion is the strongest indicator of functional disability in children with systemic JIA. Loss of joint motion has a greater effect on lower limb function.

Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in childhood and a leading cause of childhood disability¹⁻³. Most children with JIA have a satisfactory longterm disease outcome without serious disability. A small percentage, however, will enter adulthood with severe functional disabilities⁴⁻⁷.

Ten percent of children with JIA have a systemic onset, characterized by an initial phase with general disease symptoms such as spiking fever, the classic rheumatoid rash, hepato- and splenomegaly, lymphadenopathy, and pericarditis.

These systemic symptoms may precede the onset of the chronic arthritis for months. About half of children with systemic onset appear to recover completely, often after a variable period of oligoarthritic involvement. The other half continues to show progressive involvement of more and more joints and develops moderate to severe disability¹.

In children with JIA, features of active arthritis will develop. Initially, features such as swelling, hydrops, pain, and limited joint motion are prominent. In the course of time, these children can develop problems with independent ambulation, washing, dressing, and manipulative skills.

Quality of life can also be decreased as a consequence of disease activity and/or resulting impairments. Recent studies show that some children with JIA have problems in coping with their disease and social functioning⁸⁻¹⁰.

The International Classification of Impairments, Disabilities and Handicaps¹¹, based on the work of Philip Wood and developed for the World Health Organization, enables classification of the consequences of chronic illnesses. Distinctions can be made at different levels: impairments, functional disabilities, and handicaps. In JIA loss of joint motion, joint swelling, and pain are considered to be impairments due to the organic disorder. Common functional disabilities in JIA are restrictions in ambulation, washing, dressing, and manipulative skills. Handicap covers the disadvantage due to impairments or disability in social functioning. Taking personal and environmental factors into account, the model suggests a linear and causal relationship from disease to impairment, and to disability and handicap¹²⁻¹⁴.

Van der Net^{15,16} has described the relationship between impairments and functional disabilities in 4 groups of children with JIA. In these studies, impairments in joint function and disabilities in independent functioning are expressed in total scores. The correlations between impairments and disabilities appeared to be weak to moderate in the examined patient groups. In a cross sectional study of 21 children with systemic JIA¹⁵, only a moderate relationship was found between the Joint Alignment and Motion scale (JAM), which measures impairments, and the Childhood Health Assessment Questionnaire (CHAQ) as assessment tools for disability.

One of the explanations for this moderate relationship can be found in compensation. Activities with involvement of both extremities can be executed in various manners. In these activities compensatory strategies can be used to avoid involvement of the impaired joints. For example, despite serious impairment of the hip, tasks like tying shoe laces can be executed by compensatory movements in trunk and arms.

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Previous studies have not revealed the precise relationship between local joint impairments and specific disabilities because total scores of joint impairments and disabilities were used. By studying more specific activities with no possibility of compensation, it is expected that the relationship between impairments and disabilities will be more pronounced.

Physical therapy in children with JIA focuses mainly on reducing impairments (by improving joint motion, reducing joint swelling, and decreasing pain). However, patients have to deal with the associated disabilities. The key diagnostic question for treatment planning should be, which impairments that are related to patients' functional disability can also be resolved with physical therapy intervention? Insight into the precise relationship between impairments and disabilities will therefore be fundamental in goal setting for the physical therapist¹³.

We investigated the relationship between joint impairments and disabilities in children with systemic JIA.

Compared to results in other studies, we investigated this relationship at a more specific level. In particular we focused on activities with no possibility of compensation.

MATERIALS AND METHODS.

Patient selection. The data for our study were derived from a study on bone mineral density and growth in children with rheumatic diseases treated with corticosteroids from our departments of pediatrics¹⁷. During that study an extensive joint and function examination was carried out. To answer the question of our present study, the results of the physiotherapeutic examination of the children with systemic JIA were used.

The data of 21 children (14 outpatients from the Department of Pediatric Rheumatology, Sophia Children's Hospital, Rotterdam, and 7 outpatients from the Department of Pediatrics of Leiden University Medical Center) were analyzed. Eleven boys and 10 girls with systemic JIA were included. The mean age was 9.3 years (range 3.6–16.4; standard deviation, SD, 4.1). At disease onset the mean age was 4.1 years (range 1.4–8.3, SD 1.9). The mean duration of the disease was 4.8 years (range 0.8–12.6, SD 3.6).

All children had been treated with steroids for over a year. At the time of this study 12 patients still used steroids, 20 patients received nonsteroidal antiinflammatory drugs, and 17 patients used one or more second line drugs (see Table 1).

The pediatric physical therapist (WPB) examined the children for the presence of impairments in joint function and functional disabilities.

Impairment

Joint impairments were evaluated by scoring presence of capsular or joint swelling, tenderness during the joint examination, and loss of joint mobility.

Swelling. A joint count on swollen joints (JCS) was scored by using the 28 joint count according to Fuchs¹⁸, with addition of the ankle joints. The following joints were examined for swelling: the proximal interphalangeal joints (PIP), metacarpophalangeal joints (MCP), wrists, elbows, shoulders, knees, and ankles. The swelling scores for each joint were 0 (no swelling) and 1 (swelling present), resulting in a 30 joint count with a total score ranging from 0 to 30.

Tenderness (Tender Joint Count [JCT]). The following joints were examined for tenderness: shoulders, elbows, wrists, hips, knees, ankles; the remaining were evaluated as a single joint:

metacarpophalangeal (MCP), PIP, and metatarsophalangeal (MTP) joints. The tenderness scores for each joint were 0 (no tenderness) and 1 (tenderness present), resulting in a total score between 0 and 18. Joint mobility: Joint mobility was determined with a standard 2 legged goniometer and read to the nearest 5°. The limitation of joint mobility was evaluated by using the mobility scale of the Joint Alignment and Motion scale (JAM)¹⁹. The JAM scale assigns 42 movements in a limitation score, based on a determined percentage of the normal range of motion. The score for each joint varies from 0

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(normal range of motion) to 1 (between 0 and 5% decreased range of motion), 2 (between 6 and 25%), 3 (between 26 and 75%), and 4 (over 76%). The final score was determined as the sum of the left and right joints, resulting in a score between 0 and 168. The JAM also contains an alignment scale that could not be used in this study because the abnormality of posture and position in children differ from the abnormalities in the adult patient.

Disability

The extent of functional disabilities was evaluated with the Dutch version of the Childhood Health Assessment Questionnaire (CHAQ)²⁰⁻²² and the Juvenile Arthritis Functional Assessment Scale (JAFAS)²³. During this study we used the CHAQ-c questionnaire, self-reported by children between 7 and 19 years old. The parent reported CHAQ-p questionnaire was used for children younger than 7 years. The questions of the CHAQ assess independent function in activities of daily life. These activities are divided into 8 different domains: dressing and grooming, arising, walking, eating, hygiene, reach, grip, and activities. Three components are evaluated for each domain: ratings of the degree to which daily functions are difficult to perform, reported use of special aids or devices, and activities for which assistance of another person is required. Each question is scored from 0 to 3, ranging from "without any difficulty" to "unable to do." When the majority of children are not expected to be able to perform a certain activity due to age, "not applicable" is marked. The compounded CHAQ score has a possible score from 0 to 3.

The JAFAS is a standardized instrument of functional performance for children between 7 and 18 years. The JAFAS requires standardized simple equipment and can be administered in about 10 min by a health professional who clocks the child's performance on 10 physical tasks. The recorded time compared with a standard time presents the test score. The JAFAS items are scored 0 "task performed within the standard time," 1 "task performed in more than the standard time," and 2 "unable to perform the task," resulting in a possible score from 0 to 20. Discomfort: Discomfort was determined by the presence of pain and its severity in the past week, rated on a double anchored horizontal visual analog scale (Pain- VAS) with anchors of "no pain" and "very severe pain," and is included in the CHAQ^{20,21}.

Analysis. The level of joint impairments was subdivided according to extremity: arm/hand or leg. Joint motion, swelling, and tenderness were determined in the extremities. For leg impairment, joint motion was measured and number of tender (hip, knee, ankle, and MTP) and swollen (knee and ankle) joints was counted. For arm/hand impairment, joint motion was measured and number of tender and swollen joints (shoulder, elbow, wrist, MCP and PIP joints) was counted.

At the level of functional disabilities a selection of items was made for testing specific extremity function. To reduce the effect of compensation in carrying out a function, only those items were selected that exclusively test the specific extremity function. The selection of these items took place as follows: 4 physical therapists experienced in rheumatology independently selected items in CHAQ and JAFAS that test specific arm/hand or leg function.

If, according to the physical therapists, more than one extremity was involved in the performance of an activity, the item was excluded. When 3 of the therapists agreed that the activity tested one extremity only, the item was included. These items together give 4 compounded measures: the CHAQ-arm, JAFAS-arm, CHAQ-leg, and JAFAS-leg (the selected items are shown in Tables 4 and 5).

The analysis of the relationship between joint impairments and functional disabilities consisted of 3 steps: (1) The correlation between joint impairments (swelling, tenderness, and the limitation of joint motion) and functional disabilities (CHAQ, JAFAS) was calculated. (2) This correlation was calculated on the extremity level (impairment and the disabilities were therefore subdivided according to the specific extremity of interest). (3) This correlation was calculated on the joint and item level. The previously selected items were correlated with larger joints of a specific extremity. A compounded measure for mobility of each individual joint was used. For each joint, the full range of joint motion from flexion to extension for the left and right side was counted.

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For analysis at the level of the extremities and joints, only items selected in advance from the questionnaire and the function test were used.

RESULTS.

The clinical characteristics of the patients with respect to impairments and disabilities are represented in Table 1. All variables were tested for normal distribution by examining box plots and computing the kurtosis and skewness. The results of “tenderness during joint examination” and some of the derived scores (JAM-leg, swelling-leg) did not show a normal distribution. For that reason the Spearman rank correlation was used in all the analyses. The definitions we utilized to describe the relationship are in accordance with Portney²⁴. Correlations ranging from 0.00 to 0.25 indicate little or no relationship, those from 0.25 to 0.50 suggest a fair degree of relationship, values of 0.50 to 0.75 are moderate, and above 0.75 are considered good to excellent.

Total scores. The relationship between impairments and disabilities using complete instruments is represented in Table 2. The relationship between the total scores of functional disability as measured with the CHAQ and loss of joint motion (JAM) was moderate ($r_s 0.66$, $p < 0.01$). The relationship between functional disability and swollen joint count (JCS) was fair ($r_s 0.45$, $p < 0.05$). There was no relationship between reported functional disability and tender joint count. The relationship between pain-VAS and tender joint count was fair but not significant ($r_s 0.35$, NS).

[table 1][table 2].

The relationship between the total scores of the function test (JAFAS) and the loss of joint motion (JAM) was good ($r_s 0.77$, $p < 0.01$). The relationship between the function test and the total swollen joint count was moderate. There was no relationship between function test and tender joint count.

Extremities. Table 3 represents the relationship between disabilities and impairments and the extremities. The sum of the selected items out of the questionnaire and function test was compared to joint impairment of the extremity of interest. The relationships between functional disabilities and loss of joint motion in the leg (JAM-leg) were comparable to the relationships between total scores (JAM/CHAQleg, $r_s 0.58$, $p < 0.01$; JAM/JAFAS-leg, $r_s 0.86$, $p < 0.01$).

[table 3].

The relationship between functional disabilities and loss of joint motion in the arm (JAM-arm) was moderate (JAM/CHAQ-arm, $r_s 0.52$, $p < 0.05$; JAM/JAFAS-arm, $r_s 0.53$, $p < 0.05$). The only significant relationship between the functional disabilities and the swollen joint count was between swollen joint count in the arm and the JAFAS; this relationship was moderate. There was no relationship between functional disabilities and tender joint count at the level of arm or leg function.

[table 4].

Questionnaire items and joints. Tables 4 and 5 represent the relationship between functional disabilities and impairments at the level of items from the questionnaire/function test and compounded measures of range of motion of individual joints. First we calculated the association between selected CHAQ items and compounded measures of the range of motion of the individual joints. In activities included in the domains “rising,” “walking,” “hygiene,” and “general activities” there was a moderate to good relationship between the items of the questionnaire and the compounded measure of the range of motion of the hips (r_s range 0.70 to 0.81). This relationship was also found between the compounded measure for the range of motion of the knee and ankle (r_s range 0.55 to 0.80).

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[table 5].

There was mainly a moderate relationship between activities included in the domains “dressing and grooming,” “hygiene,” “reach,” “grip,” and the compounded measures of range of motion of the shoulder and elbow (r_s range 0.55 to 0.80). There was no significant relationship between items of these domains and the compounded measure of range of motion of the wrist. Activities included in the domains “dressing and grooming,” “hygiene,” “reach,” “grip,” and the compounded measures of range of motion of the shoulder and elbow (r_s range 0.55 to 0.80). There was no significant relationship between items of these domains and the compounded measure of range of motion of the wrist. Activities included in the domain “eating” and the compounded measures of the range of motion of shoulder, elbow, and wrist showed no significant relationship.

Next we calculated the association between the selected JAFAS items and the compounded measures of range of motion of the individual joints. In leg activities a moderate to good relationship was found between the selected items and the compounded measures of range of motion of the hip, knee, or ankle (r_s range 0.57 to 0.86). This relationship was similar for each joint.

The correlation between the selected JAFAS items of the arm and the compounded measure of the shoulder was moderate (r_s range 0.60 to 0.77). Between these items and the compounded measure of range of motion of elbow and wrist only one significant and moderate relationship was found.

DISCUSSION.

We investigated the relationship between joint impairments and disabilities in children with systemic JIA. We researched this relationship at a specific level, the extremities (arm/hand and leg). Our results show that from the domain of joint impairments, loss of joint motion is the strongest indicator of functional disability and has a particularly large influence on lower limb function.

In our group of patients functional disabilities were mainly induced by loss of joint motion and to a lesser extent by the number of swollen joints, but not by the number of tender joints. The relationship between loss of joint motion and disabilities was clearly stronger than in some other studies concerning children with JIA^{15,16,25-27}. This may be caused by the severity of loss of joint motion and of the disabilities in our population. In the group of patients who participated in our study, only the children with prolonged steroid treatment were included. Clearly they experienced more restrictions in joint motion and in daily function than the children in some other studies. It might be possible that only loss of joint motion above a certain level leads to functional disabilities. Badley, *et al*²⁸ sought to determine the association of limited range of joint motion with difficulty in performance of activities in adult patients with rheumatoid arthritis (RA). They found that patients with osteoarthritis or RA had difficulty with walking and transfers to and from toilet and bed if they could not flex their knees to at least 70°.

In other studies the correlation between pain or joint swelling and functional disabilities was much stronger than in our study^{15,16,25,26} and was sometimes even stronger than the relationship between functional disabilities and the loss of joint motion. Joint tenderness has been suggested as a strong determinant of physical disability in JIA^{26,27} and in patients with early RA²⁹. These varied results indicate a complex relationship between impairment and disability, determined by factors like severity and duration of the disease. In early arthritis, pain and tenderness might be the strongest factors in predicting disability. In prolonged arthritis, pain and joint swelling might become less important. Psychosocial variables such as coping strategies and affective states^{30,31} might also affect this complex relationship.

Finally, the limited number of patients in this study and the small range of tender joints could negatively have affected the relationship between the number of tender joints and functional disability.

The relationship between the JAFAS functional performance test and the impairment in joint function appeared to be stronger than the relationship between the CHAQ questionnaire and the impairments.

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An explanation might be that questionnaire measurements reflect a subjective estimation of function, and therefore presumably reflect what is important to the patient. The patient's own judgment will be affected by personal (attitude) and environmental (family support) influences. In the comparison of observed and reported function by Health Assessment Questionnaire in adult RA patients, both under- and overestimation of the actual functional limitations have been described³². The JAFAS on the other hand is an objective measure of function, performed under fixed conditions and therefore less affected by other influences, hence the stronger relationship.

To study the relationship between impairment and disability at the level of the extremity, we examined functional activities with little or no possibilities for compensation.

The relationship between loss of joint motion in the leg and disabilities in leg activities appeared to be strong.

However, the relationship between impairments and disabilities in the arm appeared to be moderate. These results are in accordance with earlier studies among elderly patients with RA, showing a weak relationship between disabilities and upper extremity joint motion²⁸. Loss of joint motion might have a large impact on functional activities of the leg, due to the necessity of weight bearing, combined with large ranges of joint motion in activities like rising, walking, or climbing the stairs. Loss of joint motion might have a minor effect on functional activities of the arm and hand. In activities like eating, grasping, or writing, coordination plays a major role and only a small range of joint motion is necessary.

The relationship between items from the questionnaire or function test and the compounded measures for range of motion of individual joints is strongest in the hip and shoulder. The mobility of the shoulder joint appeared to be the most important factor in predicting functional disabilities in the arm. Shoulder function might be more affected by loss of joint motion than elbow or wrist function, due to the necessity of large shoulder joint motion in activities like dressing, hair care, or taking something out of a closet.

These results are comparable to Badley's results²⁸ among elderly patients with RA. She described a good relationship between the range of motion of the shoulder and activities like "bending the arm" and "reaching upwards" (dressing, washing, reaching).

The mobility of the hip joint appeared to be the factor most predictive of the existence of functional disabilities in activities such as rising, getting in and out of a car, or going to the toilet. Conversely, in activities such as walking, running, and playing, the mobility of the ankle appeared to be the most predicting factor of functional disabilities.

Treatment goals of physical therapy in children with JIA can consist of maintaining or improving joint motion, reducing joint swelling and decreasing pain on the impairment level, and training by means of various activities with the aim to reduce functional disability. Dekker, *et al*³³ determined the strength of the relationship between treatment goals and physical therapy interventions. Their study showed that the aim of physical therapy was mainly to treat impairments, thereby intending to remedy an associated disability. Several studies of the relationship between impairments and disabilities, however, show a complex relationship.

Therefore, physical therapy limited to treating impairments may be questionable.

The results of our study suggest that to achieve optimal leg function, it is important to conduct not only training involving leg activities, but also joint motion of the hip, knee, and ankle. To decrease functional disabilities in the arm and hand treatment should involve training in the activities that are impaired. On the other hand, when broader activities like dressing and washing are impaired, joint motion of the shoulder deserves extra attention.

Physical therapy should aim to increase functional health. When there is a distinct relationship between the experienced disability and the underlying impairment, treatment goals on the impairment level could be the means to achieve these goals. When there is no direct relationship, physical therapy limited to treating impairment may be questionable and physical therapy should focus on the functional disability level¹³. Further research about the degree to which musculoskeletal

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impairments are associated with functional disability is important to achieve more purposeful and effective physical therapy.

REFERENCES.

1. Cassidy JT, Petty RE. Juvenile rheumatoid arthritis. *Textbook of pediatric rheumatology*. New York: Churchill Livingstone; 1990:113-219.
2. Petty RE, Southwood TR, Baum J, et al. Revision of the proposed classification criteria for juvenile idiopathic arthritis: Durban 1997. *J Rheumatol* 1998;25:1991-4.
3. Woo P, Wedderburn LR. Juvenile chronic arthritis. *Lancet* 1998;351:969-73.
4. Wallace CA, Levinson JE. Juvenile rheumatoid arthritis: outcome and treatment for the 1990s. *Rheum Dis Clin North Am* 1991;17:891-905.
5. Ruperto N, Levinson JE, Ravelli A, et al. Longtime health outcome and quality of life in American and Italian inception cohorts of patients with juvenile rheumatoid arthritis. I. Outcome status. *J Rheumatol* 1997;24:945-51.
6. Graham BT, Lovell DJ. Outcome in pediatric rheumatic disease. *Curr Opin Rheumatol* 1997;9:434-9.
7. Petty RE. Prognosis in children with rheumatic diseases: justification for consideration of new therapies. *Rheumatology* 1999;38:739-42.
8. Huygen ACJ, Sinnema G. Psychologische en sociale aspecten van Juvenile Chronische Artritis. *Tijdschr Kindergeren* 1990;59:173-8.
9. Koopman HM, ten Cate R, van Rossum M, et al. Health related quality of life of children with juvenile chronic arthritis [abstract]. *Rev Rheum Engl Ed* 1997;10 Suppl:240S-K5.
10. Miller JJ. Psychosocial factors related to rheumatic disease in childhood. *J Rheumatol* 1993;20 Suppl 38:1-11.
11. International classification of impairments, disabilities and handicaps. Geneva, Switzerland: World Health Organisation; 1980.
12. Guccione AA. Arthritis and the process of disablement. *Phys Ther* 1994;74:408-14.
13. Guccione AA. Physical therapy diagnosis and the relationship between impairments and function. *Phys Ther* 1991;71:499-504.
14. Jette AM. Physical disablement concepts for physical therapy research and practice. *Phys Ther* 1994;74:380-6.
15. Van der Net J, Prakken ABJ, Helders PJM, et al. Correlates of disablement in systemic onset juvenile chronic arthritis: A cross sectional study. *Scand J Rheumatol* 1997;26:188-96.
16. Van der Net J, Prakken AB, Helders PJ, et al. Correlates of disablement in polyarticular juvenile chronic arthritis – a cross sectional study. *Br J Rheumatol* 1996;35:91-100.
17. Van Schrieck SCA, Mul D, van Suijlekom-Smit, et al. Bone mineral density and growth in children with rheumatic diseases treated with corticosteroids [abstract]. *Rev Rheum Engl Ed* 1997;10 Suppl:244S.
18. Fuchs HA, Brooks RH, Callahan LF, Pincus T. A simplified twentyeight- joint quantitative articular index in rheumatoid arthritis. *Arthritis Rheum* 1989;32:531-7.
19. Spiegel TM, Sinden Spiegel J, Paulus HE. The Joint Alignment and Motion scale: a simple measure of joint deformity in patients with rheumatoid arthritis. *J Rheumatol* 1987;14:887-92
20. Singh G, Athreya BH, Fries JF, Goldsmith DP. Measurement of health status in children with juvenile rheumatoid arthritis. *Arthritis Rheum* 1994;37:1761-9.
21. Huygen ACJ, de Graaf IMAB, Sinnema G. Dutch translation of the Children's Health Assessment Questionnaire. Utrecht: Wilhelmina Childrens Hospital (WKZ); 1991.
22. Len C, Goldenberg J, Bosi Ferraz M, Hilario MO, Olivera LM, Sacchetti S. Crosscultural reliability of the Childhood Health Assessment Questionnaire. *J Rheumatol* 1994;21:2349-52.

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23. Lovell DJ, Howe S, Shear E, et al. Development of a disability measurement tool for juvenile rheumatoid arthritis: The Juvenile Arthritis Functional Assessment Score. *Arthritis Rheum* 1989;32:1390-5.
24. Portney LG, Watkins MP. *Foundations of clinical research, applications to practice*. Norwalk, CT: Appleton & Lange; 1993.
25. Ravelli A, Viola S, Ruperto N, Corsi B, Ballardini G, Martini A. Correlation between conventional disease activity measures in juvenile chronic arthritis. *Ann Rheum Dis* 1997;56:197-200.
26. Flatø B, Sorskaar D, Vinje O, et al. Measuring disability in early juvenile rheumatoid arthritis: evaluation of a Norwegian version of the Childhood Health Assessment Questionnaire. *J Rheumatol* 1998;25:1851-8.
27. Ruperto N, Giannini EH. Redundancy of conventional articular response variables used in juvenile chronic arthritis trials. *Ann Rheum Dis* 1996;55:73-5.
28. Badley EM, Wagstaff S, Wood PHN. Measures of functional ability (disability) in arthritis in relation to impairment of range of joint motion. *Ann Rheum Dis* 1984;43:563-9.
29. Van Leeuwen MA, van der Heijde DMFM, van Rijswijk MH, et al. Interrelationship of outcome measures and process variables in early rheumatoid arthritis. A comparison of radiologic damage, physical disability, joint counts and acute phase reactants. *J Rheumatol* 1994;21:425-9.
30. Hagglund KJ, Haley WE, Reveille JD, Alarcón GS. Predicting individual differences in pain and functional impairment among patients with rheumatoid arthritis. *Arthritis Rheum* 1989;32:851-8.
31. Boström C, Harms-Ringdahl K, Nordemar R. Shoulder, elbow and wrist movement impairment-predictors of disability in female patients with rheumatoid arthritis. *Scand J Rehabil Med* 1997;29:223-32.
32. Van den Ende CHM, Hazes JMW, le Cessie S, Breedveld FC, Dijkmans BAC. Discordance between objective and subjective assessment of functional ability of patients with rheumatoid arthritis. *Br J Rheumatol* 1995;34:951-5.
33. Dekker J, van Baar ME, Curfs EC, Kerssens JJ. Diagnosis and treatment in physical therapy: an investigation of their relationship. *Phys Ther* 1993;73:568-80.

Bekkering, W.P., Cate, R. ten, Suijlekom-Smit, L.W.A. van, Mul, D., Velde, E.A. van der, Ende, C.H.M. van den. The relationship between impairments in joint function and disabilities in independent function in children with systemic juvenile idiopathic arthritis. *Journal of Rheumatology*: 2001, 28(5), 1099-1105

Tables

Table 1. Clinical characteristics of 21 patients with systemic juvenile idiopathic arthritis.

	N	Mean ± SD [Range (maximal score)]
General		
Boys	11	
Girls	10	
Age	—	9.3 ± 4.1 [3.6–16.4]
Duration of disease		4.8 ± 3.6 [0.8–12.6]
Age of onset		4.1 ± 1.9 [1.4–8.3]
Medication		
NSAID	20	
SAARD (Gold, Plaquenil)	5	
Salazopyrine	5	
Steroids	12	
Methotrexate, Immuran	14	
Impairment level		
JAM	21	42.0 ± 37.4 [0–119 (168)]
Swollen Joint Count (JCS)	31	10.8 ± 7.3 [0–26 (30)]
Tender Joint Count (JCT)	21	3.1 ± 4.5 [0–16 (18)]
Disability level		
CHAQ	18*	1.7 ± 0.7 [0.4–2.9 (3.0)]
Pain-VAS	17*	1.0 ± 0.8 [0–2.8 (3.0)]
JAFAS	15 [#]	5.1 ± 4.6 [0–16 (16)]

*Missing data due to incompleteness/items “not applicable” for all children.

**Missing data due to age under 7 years.

NSAID: nonsteroidal antiinflammatory drugs.

SAARD: slow acting antirheumatic drugs.

JAM: Joint Alignment and Motion Scale.

JAFAS: Juvenile Arthritis Functional Assessment Scale.

VAS: visual analog scale.

CHAQ: Childhood Health Assessment Questionnaire.

Table 2. Spearman rank correlation between the total scores of Childhood Health Assessment Questionnaire (CHAQ), Juvenile Arthritis Functional Assessment Scale (JAFAS) and the joint impairments.

	CHAQ	JAFAS
JAM	0.66**	0.77**
Swollen Joint Count (JCS)	0.45*	0.52*
Tender Joint Count (JCT)	0.028	-0.14

* $p < 0.05$; ** $p < 0.01$.

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Table 3. Spearman rank correlation between the extremity specific parts of CHAQ, JAFAS, and joint impairments.

	CHAQ-arm	JAFAS-arm	CHAQ-leg	JAFAS-leg
JAM-arm	0.52*	0.53*		
JCS-arm	0.34	0.53*		
JCT-arm	-0.004	-0.01		
JAM-leg			0.58**	0.86**
JCS-leg			0.26	0.37
JCT-leg			0.19	0.23

* $p < 0.05$; ** $p < 0.01$.

JCS: swollen joint count.

JCT: tender joint count.

JAM: Joint Alignment and Motion Scale.

CHAQ: Children's Health Assessment Questionnaire.

JAFAS: Juvenile Arthritis Functional Assessment Scale.

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Table 4. Spearman rank correlation between specific items of CHAQ, JAFAS and a compounded measure for the range of motion of shoulder, elbow and wrist, respectively. Compounded measures for the range of motion: shoulder (anteflexion left + right); elbow (flexion left + extension left + flexion right + extension right); wrist (dorsalflexion left + palmarflexion left + dorsalflexion right + palmarflexion right).

	Shoulder	Elbow	Wrist
Childhood Health Assessment Questionnaire			
A. Dressing and grooming			
1. Dressing (incl. shoelaces and buttons)	-0.62**	-0.65**	-0.25
2. Shampoo his/her hair	-0.76**	-0.35	-0.17
4. Cut fingernails	-0.32	-0.23	0.24
C. Eating			
1. Cut own meat	-0.48	-0.31	0.03
2. Lift cup or glass to mouth	-0.21	-0.17	0.10
3. Open a new pack of milk	-0.19	-0.02	0.41
E. Hygiene			
1. Wash and dry entire body	-0.73**	-0.68**	-0.31
4. Brush teeth	-0.05	-0.21	-0.13
5. Comb/brush hair	-0.69**	-0.60*	-0.45
F. Reach			
1. Heavy object from above the head	-0.48*	-0.61**	
2. Pick up from the ground	-0.67**	-0.70**	
3. Pull on sweater over the head	-0.47*	-0.43	
G. Grip			
1. Write with pen or pencil	-0.25	-0.25	-0.24
2. Open car door	-0.57*	-0.55*	-0.33
3. Open jar	-0.65**	-0.28	-0.44
4. Turn faucets on/off	-0.42	-0.56*	-0.24
5. Push open door/turn a door knob	-0.49*	-0.51*	-0.28
Juvenile Arthritis Functional Assessment Scale			
1. Button a shirt	-0.60*	-0.51	-0.09
2. Put on a sweater/shirt	-0.63*	-0.13	-0.38
3. Cut meat	-0.77**	-0.57*	-0.06

* $p < 0.05$; ** $p < 0.01$.

$n < 14$ (missing data because items were "not applicable" for all children).

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Table 5. Spearman rank correlation between specific items out of the CHAQ, JAFAS, and a compounded measure for the mobility of hip, knee and ankle respectively. Compounded measure for the range of motion: hip (flexion left + extension left + flexion right + extension right); knee (flexion left + extension left + flexion right + extension right) and ankle (dorsalflexion left + plantarflexion left + dorsalflexion right + plantarflexion right).

	Hip	Knee	Ankle
Childhood Health Assessment Questionnaire			
B. Arising			
1. From low chair or ground	-0.71**	-0.55*	-0.62*
2. Get in and out bed	-0.72**	-0.57*	-0.70**
D. Walking			
1. Outdoors on flat ground	-0.70*	-0.57*	-0.80**
2. Climb up 5 steps	-0.43	-0.46	-0.44
E. Hygiene			
2. Take a tub bath	-0.81**	-0.62*	-0.66*
3. Get on and off toilet	-0.71**	-0.59*	-0.65**
H. General activities			
1. Shopping (#)			
2. Get in/out of car	-0.73**	-0.65**	-0.59**
3. Ride bike or tricycle (#)			
4. Domestic work (#)			
5. Run and play	-0.70**	-0.62**	-0.78**
6. Gymnastics (#)			
Juvenile Arthritis Functional Assessment Scale			
1. Sit down on the ground and arise	-0.57*	-0.68**	-0.66**
2. Walk 15 m without support	-0.66**	-0.80**	-0.64**
3. Ascend 5 stairs	-0.74**	-0.65**	-0.86**

*p < 0.05; **p < 0.01.

#p < 14 (missing data because items were "not applicable" for all children).