(Pearson's correlation coefficients (r) ranging from 0.48 to 0.92 ($n=18$) and from 0.63 to 0.88 ($n=13$), respectively, $p<0.05$). Patients with JA had lower levels of physical wellbeing (mean physical score (PhS) 41.7 v 56.0, $p<0.001$) and emotional and social wellbeing (mean psychosocial summary score (PsS) 50.4 v 55.7, $p<0.001$) than controls. At baseline, the CHQ-pf50 physical score (PhS) correlated with the child HAQ ($r=-0.52$, $p<0.001$), patients' assessment of pain ($r=-0.61$, $p<0.001$), fatigue ($r=-0.48$, $p<0.001$), global assessment of wellbeing ($r=-0.60$, $p<0.001$), physicians' global assessment of disease activity ($r=-0.47$, $p<0.001$), and morning stiffness ($r=-0.22$, $p<0.05$). Pain ($\beta = -0.40$, $p<0.001$) and physicians' global assessment of disease activity ($\beta = -4.9$, $p<0.001$) were the most important correlates of PhS in the multiple regression analysis. From baseline to follow up there was an improvement in the mean PhS, HAQ, and physicians' global assessment of disease activity (3.3, -0.18, and -0.60, respectively, $p<0.01$). The change in PhS correlated with the change in HAQ ($r=-0.46$, $p<0.001$) and in the physicians' global assessment of disease activity ($r=-0.38$, $p<0.01$).

Conclusion—The Norwegian version of the CHQ is a reliable and valid instrument for measuring physical wellbeing in children with early JA.

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4.18 Exploration of different clinical measures used in juvenile idiopathic arthritis

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Background—In 1998 the Paediatric Rheumatology International Trials Organisation (PRINTO) formulated a core set of outcome variables. They include among others: the number of joints with active arthritis, limited range of motion, and functional ability. For each domain several measures are available.

Objective—To explore the relation between these different measures.

Methods—Twenty one children with systemic onset juvenile idiopathic arthritis (JIA) with an average age of 9.3 years (SD 4.1) and mean disease duration of 4.8 years (SD 3.6) participated in this study. The number of active joints was determined by a tender joint count and a swollen joint count. Limited range of motion was determined by the JAM and the Paediatric EPM-ROM scale. Disability was determined by the performance test JAFAS and the questionnaires CHAQ, and JAFAR.

Results—The strongest relation was found between the two measures of joint motion ($r=0.94$, $p>0.001$). A weaker relation was found between the three measures of disability ($r=0.70-0.79$, $p>0.05$). No relation was found between the joint count measures. The relation between impairment and disability was the strongest in the joint motion measures compared with the JAFAS performance test ($r=0.73-0.77$, $p>0.05$) or CHAQ questionnaire ($r=0.66-0.67$, $p>0.05$).

Conclusion—Strong relations were found between scores within the same domain. Except for the joint count measures, data from different instruments concerning the same

aspect appeared to be exchangeable. Weaker relations were found between measures on different domains.

4.19 An international survey in 29 countries on health related quality of life (HRQOL) in juvenile idiopathic arthritis (JIA). Part I: a disease specific instrument: the Childhood Health Assessment Questionnaire (CHAQ)

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Objective—To evaluate HRQOL in 1492 children with JIA and 1576 healthy children as measured by the parent version of the CHAQ. CHAQ is a validated disease-specific instrument measuring functional ability in 8 daily living activities averaged in the Disability Index (DI) score ranging from 0 to 3 with higher scores indicating worst disability, and two 10 cm visual analogue scales (VAS) for pain (pain VAS) and overall wellbeing (parent VAS) assessment.

Methods—The CHAQ has been cross culturally adapted in all countries according to standard guidelines: 1–3 forward/backward translations, pretesting, and data collection on healthy children and children with JIA. Data from all countries were combined. Comparisons between groups were made by analysis of variance.

Results—Values in brackets are medians. 3068 children were enrolled from 29 countries: 1576 were healthy (age 11.2 years), 303 systemic (sys) (age 9.3), 532 polyarticular (poly) (age 10.5), 328 extended oligoarticular (ext-oli) (age 9.4), and 329 oligoarticular (oligo) (age 8.7). There was no difference in disease duration for the 4 JIA types ($p=0.12$). CHAQ results: healthy (DI=0, pain VAS=0, parent VAS=0), sys (DI=1.1, pain VAS=2.4, parent VAS=2.4), poly (DI=1, pain VAS=2.8, parent VAS=3.5), ext-oli (DI=0.6, pain VAS=2.4, parent VAS=2.1), oligo (DI=0.1, pain VAS=0.5, parent VAS=0.5). Comparisons of healthy children with sys, poly, ext-oli, oligo showed consistently a $p<0.0001$ for DI, pain VAS, parent VAS, and all the 8 domains of the CHAQ. **Conclusions**—The cross culturally adapted versions of CHAQ can discriminate between the different JIA types and healthy children. Patients with sys, poly, and ext-oli JIA have the worst functional ability and were similar to each other while patients with oligo JIA were comparable with healthy children.

4.20 Part II: A generic instrument: the Child Health Questionnaire (CHQ)

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Objective—To evaluate HRQOL in 1492 children with JIA and 1576 healthy children as measured by the parent version of the CHQ.

CHQ is a validated generic instrument measuring 14 health concepts ranging from 0 to 100, with a higher score indicating better wellbeing. 10/14 health concepts are then grouped in 2 summary scores ranging from 0 to 70, with a higher score indicating better wellbeing: the physical (PhS), and the psychosocial summary score (PsS).

Methods—CHQ has been cross culturally adapted in all countries according to standard guidelines: 1–3 forward/backward translations, pretesting, and data collection. Data from all countries were combined. Comparisons between groups were made by analysis of variance.

Results—Values in brackets are medians. 3068 children were enrolled: 1576 were healthy (age 11.2 years), 303 systemic (sys) (age 9.3), 532 polyarticular (poly) (age 10.5), 328 extended oligoarticular (ext-oli) (age 9.4), and 329 oligoarticular (oligo) (age 8.7). There was no difference in disease duration for the 4 JIA types ($p=0.12$). The CHQ results were: healthy (PhS=57, PsS=53), sys (PhS=33, PsS=46), poly (PhS=35, PsS=48), ext-oli (PhS=38, PsS=47), oligo (PhS=49, PsS=52). Comparisons of healthy children with children with sys, poly, ext-oli, oligo JIA showed consistently a $p<0.0001$ for PhS and PsS and for the 14 CHQ health concepts. Erythrocyte sedimentation rate, physicians' evaluation of disease activity on a 10 cm VAS, number of swollen joints, number of joints with pain/tenderness, number of joints with limited motion, and number of active joints were different in the 4 JIA types ($p<0.0001$).

Conclusions—The cross culturally adapted versions of the CHQ can discriminate between the different JIA types and healthy children. Children with sys, poly, and ext-oli JIA have the worst HRQOL and were similar to each other, whereas children with oligo JIA were comparable with healthy children.

5 Infection and arthritis

5.1 Hepatitis B vaccination in juvenile idiopathic arthritis

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Objective—To evaluate the responsiveness to hepatitis B vaccination in children with juvenile idiopathic arthritis (JIA) and to determine a useful vaccination schedule.

Patients and methods—39 (18 female, 21 male) children with JIA who were all in remission and HBsAg negative were enrolled into the study group. The control group consisted of 41 healthy children (21 female, 20 male). The children were divided into two groups and given two different vaccination schedules. The first group were vaccinated at 0, 1, and 3 months while the children in the second group were vaccinated at 0, 1, and 6 months respectively. Those weighing above 20 kg were vaccinated with a 20 µg dose of DNA recombinant vaccine and those below 20 kg were given 10 µg. Positive responsiveness to the vaccine was defined as an anti-HBs titre >10 mIU/ml.

Results—All children with the exception of one child with systemic JIA developed antibody response. None of the patients with JIA had a flare up or clinical deterioration related to the vaccination. There was no statistical difference of antibody levels between

the subgroups of patients with JIA. The mean (SD) antibody levels in the children with JIA were significantly lower (175.5 (118.4) mIU/ml) than those of the healthy subjects (317.08 (180.5) mIU/ml). There was no difference between the sexes in vaccine responsiveness. When the antibody levels between the two vaccination schedules were compared in healthy children, there was no statistical difference. However, there was a slightly higher, but statistically insignificant, response in JIA subjects vaccinated in group II than in those in group I. The vaccine responsiveness was not influenced either by methotrexate or prednisolone treatment. However, there was a negative correlation between prednisolone dose and anti-HBs titre ($r = -0.23$).

Conclusion—Children with JIA responded adequately to hepatitis B vaccination, and this response was not negatively influenced by immunosuppressive treatment. The more appropriate vaccination schedule for children with JIA is the schedule given at 0, 1, and 6 months.

5.2 Prevalence of HLA class II in Sydenham chorea

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Background—The relation between acute rheumatic fever (ARF) and human leucocyte antigens (HLA) is available for rheumatic heart disease. Susceptibility to Sydenham chorea (SC), a major manifestation of ARF, has been associated with the D8/17 antigen present in B lymphocytes, but data about HLA class II antigens are scarce.

Objective—To study the prevalence of HLA class II antigens among patients with SC. This was the first part of a prospective study to obtain data about HLA class I and II in patients with the different major manifestations of ARF.

Patients and methods—Patients with SC, isolated or in combination with other major manifestations of ARF, seen in our paediatric rheumatology units between 1989 and 1998, were tested for HLA class II antigens by the polymerase chain reaction with sequence-specific primers technique, with genomic DNA extracted from peripheral blood with EDTA. The frequency of HLA class II alleles was compared with that of 85 healthy volunteers using the χ^2 and/or the Fischer's exact test.

30 patients (17 female, 13 male) were enrolled in the study—6 with isolated chorea and 24 with chorea and other major criteria. The mean age at diagnosis was 14.3 years and mean follow up was 45 months. A negative association with HLA-DR8 ($p < 0.03$) was seen, and 4 of the 6 patients with chorea alone presented HLA-DR1 (χ^2 with Mantel-Haenszel = 7.61; $p < 0.006$).

Conclusions—Although the number of patients studied up to now is small, we conclude that there seems to be a gene in the HLA region which may be related to susceptibility or protection for SC.

5.3 Incidence of rheumatic fever in children in Latvia

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Background—During the past five years the incidence of acute rheumatic fever has increased.

Methods—A retrospective case history analysis over the period 1994–2000 was carried out using John's criteria, including incidence, sex, age, progression and outcome of illness. **Results**—107 patients were studied. 11 (10%) were aged under 7 years, 92 (86%) were aged 7–16 years, 4 (4%) were aged 16–18 years. 49 (46%) were female and 58 (54%) were male. In 44 (41%) patients the illness was accompanied by fever, in 46 (43%) by an increased C reactive protein level, in 47 (43%) by leucocytosis, in 52 (49%) by an increased erythrocyte sedimentation rate (> 40 mm/1st h), in 68 (64%) by antistreptolysin O titres (from 500 to 3000 IU/l). In 61 (57%) patients arthralgia was detected and poly/monarthrit in 56 (53%). Endocarditis was detected in 60 (56%) patients, damage of the mitral valve in 34 (31%), of the aortic valve in 16 (15%), and of both the mitral and aortic valves in 10 (9%). In 5 (5%) cases pancarditis and in 84 (79%) cases myocarditis were detected. In 6 (6%) cases chorea minor was seen. A relapse of the rheumatic fever was seen in 13 (12%) children.

Conclusions—The current study underlines the fact that the severity and incidence of rheumatic fever in Latvia have increased during the past 5 years. The illness more often proceeds with endocarditis (damage of the mitral or combined mitral/aortic valves) and has reached a level of 5.1/100 000. The risk group is school age children.

5.4 PANDAS case in Latvian girl

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Background—PANDAS (paediatric autoimmune neuropsychiatric disease associated with streptococci) is an acronym describing neuropsychiatric symptoms resulting from autoimmune responses to streptococcal (group A β haemolytic) infection. The diagnostic criteria for PANDAS are (a) the presence of an obsessive compulsive disorder and a tic disorder; (b) paediatric onset of symptoms (age 3 years to puberty); (c) an episodic course of symptom severity; (d) association with group A β haemolytic streptococcal infection; and (e) association with neurological abnormalities.

Objective—To describe cases of rare syndromes related to rheumatic fever.

Results—An adolescent girl aged 15 who had arthritis of both knees and feet suddenly developed emotional lability, cognitive deficits, oppositional behaviours, and motor hyperactivity. Investigation showed mitral valvulitis, regurgitation, a raised erythrocyte sedimentation rate, and an antistreptococcal antibody titre. During a six month follow up the heart disease appeared and the obsessive compulsive and tic disorders improved.

Conclusion—The PANDAS case corresponds with described diagnostic criteria (Kleinsasser, 1999, Schulman, 1999).

5.5 Juvenile spondyloarthropathy associated with *Mycoplasma pneumoniae* infection

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Objective—Reactive arthritis (ReA) has been sporadically reported as triggered by *M pneumoniae*. This study examined the potential relation between acute *M pneumoniae* infection and juvenile spondyloarthropathy (jSpA). **Patients and methods**—Seven patients (4 female, 3 male) with jSpA secondary to *M pneumoniae* were examined. Their mean age at onset was 8.3 years (range 2–13), and the mean duration of follow up was 5.5 months (range 2.5–11). All patients had < 4 joints affected. *M pneumoniae*-specific IgM, IgG, and IgA antibodies were serologically confirmed by enzyme linked immunosorbent assay (ELISA) tests (Savyon Diagnost, Israel). Owing to the early appearance and relatively short lifetime of *M pneumoniae*-specific IgM antibodies, their detection allowed the diagnosis of acute infection using a single serum sample, confirmed by a parallel serum in 4/7 patients. Immunoglobulin concentrations higher than 10 U/l are considered positive and those higher than 50 U/l as highly positive. Specific IgA antibodies were detected in only patient.

Results—The mean recovery time was 2.5 weeks (range 1–4) in 5 patients with ReA; one patient developed juvenile rheumatoid arthritis (pauci-type II) and in one case juvenile ankylosing spondylitis was diagnosed. Two patients were HLA-B27 positive, and one patient was HLA-B7 positive. Four patients had preceding respiratory symptoms, and only two were treated with antibiotics.

Conclusion—These findings provide evidence of jSpA after an acute *M pneumoniae* infection. Our results suggest that detecting *M pneumoniae*-specific antibodies in serological screening of patients with jSpA might be useful. It is presently unclear whether antibiotic treatment would change the disease course in such patients.

6 Juvenile dermatomyositis

6.1 A preliminary comparative study of high frequency muscle ultrasound and magnetic resonance imaging in 7 patients with juvenile dermatomyositis

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Objective—To assess the value of ultrasound scans (USS) of the thigh in juvenile dermatomyositis (JDM) in comparison with magnetic resonance imaging (MRI). To correlate these findings with clinicopathological indicators of disease activity.

Methods—7 white patients with JDM (4 male, 3 female) had a high resolution muscle ultrasound examination. Mean age at onset was 6.8 years (range 1–15), 5/7 were scanned at diagnosis (within 6 months of diagnosis) and at follow up, 2/7 were scanned owing to chronic disease at 6 years from disease onset. Results of these scans were correlated with clinical, biochemical and histological markers. USS were correlated with MRI of the thigh in all patients.

Results—All 5 USS performed at diagnosis were abnormal. Most showed diffuse symmetrical hyperechogenicity affecting mainly