showed that the presence of damage was positively associated with the SLEDAI score (odds ratio (OR)=1.1, $p=0.005$), cumulative drug score (OR=1.0, $p<0.001$), and NPS disease at onset (OR=4.4, $p=0.008$) and negatively associated with Raynaud phenomenon at onset (OR=0.2, $p=0.01$).

Conclusions—Our results show that permanent damage is common in paediatric onset SLE and is significantly associated with lupus disease activity and cumulative drug treatment. NPS disease at onset is a strong predictor of late damage, whereas the presence of Raynaud phenomenon may exert a protective effect.

3.9 Uncommon causes of liver disease in juvenile systemic lupus erythematosus

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Background—Severe liver disorders are rare in patients with systemic lupus erythematosus (SLE). We report two patients with juvenile SLE who developed uncommon hepatic complications.

Case reports—Patient No 1: A 10 year old girl was admitted with a 3 week history of malar rash, intermittent arthritis, fatigue, and abdominal pain. Investigations showed haemolytic anaemia, thrombocytopenia, and positive antinuclear antibodies (ANA) and anti-DNA antibodies. High titre antinuclear antibodies (aCL) and lupus anticoagulant (LA) were detected. A diagnosis of SLE was established. During a stay in hospital the girl developed enlargement of the liver, which was tender at palpation. Ultrasonography showed a large thrombus occluding the lumen of the inferior vena cava (Budd-Chiari syndrome). Based on the association of vascular thrombosis and positive antiphospholipid antibodies, antiphospholipid syndrome was diagnosed and long term anticoagulation treatment was started.

Patient No 2: A 9 year old girl, who had had autoimmune thyroiditis since the age of 6 years, was admitted with a 1 week history of fever, fatigue, arthralgia, and purpuric rash in the legs. Physical examinations showed hepatosplenomegaly, ascites, and oedema of the ankles. Laboratory investigations disclosed pancytopenia, liver transaminase increase, hypoalbuminaemia, low serum complement levels, and positive ANA and anti-DNA antibodies. aCL and LA were absent. Liver ultrasonography showed an enlarged liver with diffuse pseudonodular changes. A percutaneous biopsy showed necrotising arteritis with nodular regenerative hyperplasia of the liver. A diagnosis of SLE with necrotising arteritis of the liver was made, and treatment with high dose prednisone and cyclophosphamide was begun.

Conclusion—The reported patients demonstrate the protean clinical spectrum of liver disease in juvenile SLE.

4 Epidemiology and outcome

4.1 Epidemiology of rheumatological diseases in Latvia

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Objective—To research the epidemiology of rheumatic diseases in children and adolescents aged 1–18 years to obtain comparative data with those of other countries.

Methods—We used the children's rheumatic diseases register data obtained by 1 January 2000 to determine incidence and prevalence.

Results—1251 children and adolescents aged 1–18 have rheumatic diseases. 70.4% have been diagnosed juvenile chronic arthritis (JCA), with prevalence 165.1/100 000 and incidence 34.3/100 000. 36% have registered oligoarthritis, 61% polyarthritis (rheumatoid factor positive 5.2%), 2% systemic form arthritis, 0.5% psoriasis with arthritis, 0.5% Crohn's disease and ulcerous colitis with arthritis. Eye injuries in patients with JCA occur in 2.4% of cases. The prevalence of collagenoses is 11.1/100 000 and incidence 2.1/100 000. Recent years show an increase of scleroderma with a prevalence 4.5/100 000. Since 1993 cases of rheumatic fever have increased, reaching a prevalence of 12.6/100 000 and an incidence of 2.1/100 000 in 1999. 49.3% have endocarditis. JCA and collagenoses affect mostly female children, rheumatic fever occurs mostly in male children. Patients with acute arthritis have an incidence of 10.3/100 000. Epstein-Barr virus (9.8%) and streptococcal infection (8.6%) are most common.

Conclusion—Epidemiological data in children and adolescents with JCA and collagenoses are similar to those of the developed countries, except for a smaller occurrence of eye injuries in patients with JCA. Rheumatic fever with frequent valvular injuries has a comparatively higher occurrence. In 38% of patients the cause of acute arthritis is unknown.

4.2 High prevalence of childhood chronic arthritis among the Shipibo people of Amazonian Peru

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Few prevalence figures of childhood chronic arthritis exist for developing countries and even fewer for the indigenous population living a traditional life in those countries. We undertook a prospective population based study of the prevalence rate of childhood arthritis among the Shipibo people living in the Amazonian part of Peru.

Methods—As most adults and children are illiterate and time is vaguely defined as the number of elapsed rainy or dry seasons, no established criteria could be used. Instead a questionnaire was constructed with questions that aimed at differentiating between joint disorders of traumatic, infectious, and rheumatic aetiology. The questionnaire was given orally by an interpreter to all children present in the villages at time of the investigation and to a close adult relative. All children were physically examined for the presence of arthritis. The study area was Shipibo villages accessible from a smaller town, and the study was performed during January and February 1999.

Results—651 of 785 children (83%) living in 6 areas of Yarina Cocha and 2 villages were examined personally by two of us and had the questionnaire applied. 4.3% had joint complaints of traumatic cause, 1.4% of possible infectious origin, and 1.8% a possible rheumatic joint disorder. Of the 12 children with a possible chronic arthritis, 4 had active arthritis at time of investigation, which corresponds with a prevalence of 614 per 100 000 children <16 years of age. If all 12 children with a possible arthritis are included the prevalence is 1843 per 100 000 children <16 years of age. This very high prevalence must

be viewed with caution as the study period was short, the most remote part of the Shipibo area could not be accessed, the concept of medicine is traditional, and longstanding reactive arthritis might have been included. This means that the prevalence as well as possibly being overestimated might even have been underestimated. Also among adults of the Shipibo people a very high prevalence, close to 10% has been reported, indicating that the prevalence of chronic arthritis among children also may be high.

4.3 Adaption and application of child health assessment questionnaire (CHAQ) to an indigenous population of Amazonian Peru

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The CHAQ has been successfully adapted and applied to most industrialised societies and also to a country in transition like Costa Rica. Common to all those countries is a high educational level. We attempted to determine whether it was possible to adjust the Costa Rican version of the CHAQ (CR-CHAQ) to the conditions and language of the Shipibo people, a well defined indigenous group living a traditional life in the Amazonian part of Peru.

Method—A cultural adaptation and linguistic translation of certain key words of the CR-CHAQ was done. It was tested on children and parents without joint disorders and was with minor alterations well understood. Self administration was not possible, however, as most adults and children cannot read or write. Instead an interpreter gave the Shipibo-CHAQ (SH-CHAQ). The SH-CHAQ was tested on 12 children with possible rheumatic joint disorders, and a close adult. For children below 10 years of age, the SH-CHAQ was applied only to the adult.

Results—Cross cultural validity: Most questions had to be adapted to Shipibo lifestyle and traditions. The children gave higher disability scores than the adults. Test-retest: A high correlation was found between the disability index for the first and second application both for children and adults. The correlation for children (Spearman $r_s=0.994$, $p<0.002$) was higher than that for the adults ($r_s=0.839$, $p<0.02$). Interobserver score correlation: No correlation was found between the disability index for children and adults at the first application, but the second application showed a good correlation ($r_s=0.975$, $p<0.02$). The difference between the first and second application might be due to exaggeration by parents who expected to obtain health care if the child was more severely ill.

Conclusion—By carefully analysing the CHAQ and adapting the questions to the Shipibo culture we could successfully apply the new SH-CHAQ with a high test-retest and interobserver reliability.

4.4 Remission in juvenile chronic arthritis: a cohort study of 683 consecutive cases

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Evaluation of the prognosis of the juvenile forms of chronic arthritides is difficult because most patients are lost at follow up when they reach adulthood. Our institute is a privileged observatory as it is open to both juvenile and adult patients, so that children do

not need to change facility when they grow up and their records can be gathered without interruption. The files of 683 consecutive patients (463 F, 220 M) affected by juvenile chronic arthritis (JCA) attending our centre since 1970 were reviewed to establish the frequency of remission in this disease. Remission was defined as a lack of signs of disease activity (active joints and/or laboratory positive tests) in the absence of antirheumatic treatment for at least 6 months. Our series was composed of 420 cases of pauci onset juvenile rheumatoid arthritis (JRA), 88 of systemic onset JRA, 108 of poly onset JRA, and 67 of juvenile spondyloarthropathies. The patients were aged 2–41 years at the last visit with a mean period of follow up of 10.2 years (range 1–36). The remission rate was calculated taking into account five main variables: the sex, the clinical form, the age at disease onset, the duration of the follow up, and the delay in joining our tertiary care centre. With regard to disease course the pauci-JRA and the juvenile spondyloarthropathies had the highest remission rate (44% for both groups), for the systemic form it was intermediate (33%), and the poly group had the lowest remission rate (22%). None of the rheumatoid factor positive patients with poly-JRA ever went in remission. The remission rate was scarcely influenced by the sex, by the duration of the follow up, and by the age at disease onset, whereas it seemed to be influenced by the delay in joining the tertiary care centre.

4.5 Hospital admissions of rheumatic fever in Uberlândia, Brazil

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Rheumatic fever usually occurs in populations that live in poor conditions and receive inadequate primary health care. Uberlândia is a Brazilian city that has around 500 000 inhabitants who rely on good economic and public health indicators. We aimed at verifying the epidemic overview of rheumatic fever in the city and region by studying admissions to the local University Hospital between 1988 and 1998. The demographic and clinic data of the patients under 18 years old admitted with rheumatic fever and diagnosed according to the Jones criteria were analysed. A hundred and fifty nine admissions (80 (50%) male), average age 125 months (range 58–209) satisfied the criteria for inclusion. Thirty one (19%) admissions were recurrences and three children (1.8%) were submitted to a valve reconstruction. A hundred and six (67%) came from the urban area of Uberlândia and 8 (5%) from the rural areas. The average admissions/year were 14, ranging from 9 outbreaks in 1988 and 24 in 1992. Carditis occurred in 117 (74%) admissions, arthritis in 85 (53%), chorea in 67 (42%), erythema marginatum in 5 (3%) and subcutaneous nodules in 4 (2%). Despite possessing a satisfactory public health care network, the epidemic situation of rheumatic fever in the region of Uberlândia is unfavourable: the hospital admissions did not decrease and a high level of recurrences and carditis was observed. Effective public health educational policies and programmes aiming at primary and secondary prophylaxis of this disease need to be implemented in the basic healthcare units.

4.6 Evaluation of health outcome instruments in Turkish children with juvenile idiopathic arthritis

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Several health outcome instruments (HOI) have been defined in juvenile idiopathic arthritis (JIA) in order to evaluate both disease activity and disease course. The most commonly used HOIs are JAFAS, JAFAR, and CHAQ. The purpose of this study is to test the reliability of these above mentioned tests in Turkish children.

Forty six children (22 female, 24 male) with JIA were enrolled into the study group. JAFAR, JAFAS, and CHAQ questionnaires were first translated into Turkish and later these translations were checked by a professional English translator. Also, Steinbrocker score (StS), visual analogue scale (VAS), and physician global assessment scale (PGAS) were used in all subjects. After all the mean ratios of the tests had been calculated, the relation between them was analysed. All patients were questioned by a single observer. The answers taken from both children and parents at the same time were defined as common answers.

Table 1 shows the mean scores derived from the tests.

Table 1

StS	CHAQ	JAFAR	JAFAS	VAS	PGAS
1.43	0.42	0.29	0.29	0.27	0.82

The scores in CHAQ test were significantly correlated with the StS ($r=0.81$), JAFAR ($r=0.86$), JAFAS ($r=0.84$), VAS ($r=0.54$), and PGAS ($r=0.73$) scoring systems respectively. The most important problem encountered during the tests was the adaptation difficulty to certain questions ($n=18$, 46%). This problem was more prominent in the JAFAR and JAFAS tests than in the CHAQ test.

In conclusion, all HOIs, especially CHAQ, are appropriate tools for JIA. However, these questionnaires, which have been defined for developed countries, have to be adapted to the conditions of developing countries.

4.7 HLA markers and prediction of clinical manifestation and outcome in systemic onset juvenile chronic arthritis

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The clinical course of systemic onset juvenile chronic arthritis (sJCA) differs among individual patients.

In this study we analysed the effect of HLA-DR on disease progression, extra-articular manifestations, the development of amyloidosis, and genetic predisposition among 43 patients with sJCA followed up for 19 (SD 7) years. The main outcome measures were radiological changes (Steinbrocker criteria), functional disability, and amyloidosis confirmed by biopsy. Each patient was also graded for the severity of systemic disease over the disease course.²

Results—The frequencies of DR4 and DR3 were found to be significantly higher in the patients' group than in controls (55.3% v 23.6%; RR=4.0, $p<0.0001$ and 48.9% v

23.6%; RR=3.1, $p<0.008$). A significant increase in the incidence of DR4 was seen in the patients subgroup with radiological evidence of joint destruction in comparison with the controls (73.7% v 23.6%, $p<0.0001$) and patients without such changes (73.7% v 25.0%, $p<0.05$). An increase in frequency of DR3 was found in patients with severe systemic symptoms persisting or recurring for more than 2 years when compared with controls (76.2% v 23.6%, $p<0.0001$) and patients with systemic features for only up to 2 years (76.2% v 22.7%, $p<0.001$). No correlation between HLA antigen frequencies and functional status or amyloidosis was established.

Conclusions—Although the contribution of HLA typing to establishing a diagnosis of sJCA is limited, HLA typing may be of value for predicting joint destruction and severity of systemic manifestation in sJCA.

1 Hochberg, *et al*, 1992.

2 Bedford, *et al*, 1992.

4.8 Continuously high incidence of chronic arthritis in northern Norway 1985–99: comparison of retrospective and prospective studies

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Epidemiological studies differ in data regarding the incidence of juvenile chronic arthritis (JCA) partly owing to differences in diagnostic criteria, patient retrieval, and study designs, but genetic predisposition and environmental factors may also be involved in these differences.

We retrospectively investigated the incidence of JCA in the total population from northern Norway (Troms and Finnmark counties, mean child population 48 488 children aged <16 years) in the years 1985–94. Cases were identified from the hospital files of the University Hospital of Tromsø, the only hospital in the study area treating JCA, using the European League Against Rheumatism criteria for JCA. From 1995 cases from the same area were prospectively studied, from 1997 as part of a multicentre study from the Nordic Study Group of Paediatric Rheumatology (NSGPR).

During the study period 162 new cases of JCA were found. The average annual incidence of JCA was found to be 22.1 per 100 000 children under 16 years of age (28.9 in girls, 16.0 in boys). The onset types of JCA were 52.5% oligoarticular, 37.0% polyarticular, 3.7% systemic, 4.9% juvenile psoriatic arthropathies, and 1.2% juvenile ankylosing arthropathies. The retrospectively obtained data from the first 10 year period (incidence 22.6 per 100 000) did not differ significantly from the prospectively obtained data from the last 5 year period (20.1 per 100 000).

The high incidence of JCA in the northernmost part of Norway as compared with most other studies needs further investigation.

4.9 Growth delay in childhood connective tissue diseases

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Background—Impairment of linear growth is one of the most common complications of connective tissue diseases in children. There are many causes, including age at onset, disease duration and activity, decreased food intake, corticosteroid treatment, and hormonal impairment.

Objective—Our study aimed at assessing the relevance of these factors in determining growth delay.

Patients and methods—Standard deviation for height (HtSDS) was evaluated in 110 patients (23 M, 87 F), mean (SD) age 13.1 (4.2) years, affected with connective tissue diseases: 30 with systemic lupus erythematosus (SLE), 11 dermatomyositis (DM), 20 systemic onset juvenile idiopathic arthritis (JIA), 19 polyarticular JIA, 16 pauci-extended JIA, and 14 pauciarticular JIA.

Results—Twenty one patients (19%) showed growth impairment (HtSDS <-2): 7 SLE, 1 DM, 7 systemic onset JIA, 3 pauci-extended JIA, 3 polyarticular JIA; no growth retardation was found in the pauciarticular group. There was no significant difference in age at onset and disease duration between patients with and without growth delay, nor in the total steroid dose. A significant difference was found between the two groups in the mean steroid dose/kg/day ($p < 0.005$). As expected, growth retardation was more common in patients with prolonged disease activity (assessed by haemoglobin, erythrocyte sedimentation rate, and C reactive protein values and, if articular disease was present, the number of active joints) and, in JIA, a high Steinbrocker functional class ($p = 0.005$). In patients with JIA, systemic onset subtype was related to a higher incidence of growth retardation: 35% of patients with systemic onset showed growth impairment. Of the patients with JIA and growth retardation, the disease in more than 50% had a systemic onset. No significant reduction was found in IGF1 and IGFBP3 values in patients with growth delay. In our experience, the main determinant of growth impairment seems to be the type of disease, in particular, systemic onset JIA, owing to more active disease and prolonged steroid treatment.

4.10 The ECLAM may be more responsive to clinical change than the SLEDAI and can be used to predict damage in childhood onset systemic lupus erythematosus (cSLE)

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Background—The reversible inflammation or disease activity (DA) in cSLE can be measured by the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI). The European Consensus Lupus Activity Measure (ECLAM) has not been validated for use in cSLE. We have shown that damage in cSLE can be measured by the Systemic Lupus International Collaboration Clinics/American College of Rheumatology Damage Index (SLICC), and that accumulated DA measured by area under the curve (AUC) of the SLEDAI is a predictor for damage. It is unknown how the ECLAM relates to damage in cSLE.

Objectives—(a) To investigate whether the ECLAM is sensitive to change in DA, and hence suitable for use in cSLE. (b) To show that accumulated DA, as measured by the AUC of the ECLAM scores is related to the damage scored by the SLICC.

Methods—Ongoing DA (ECLAM, SLEDAI) and damage (SLICC) was measured in all 66 patients with newly diagnosed cSLE between January 1992 and January 1999. The DA at the time of diagnosis (D_{oc}), 6 months after

diagnosis (D_{6m}), at the time of the 1st flare (F), and 6 months after the 1st flare (F_6) was used to estimate the sensitivity to change—measured by the effect size (ES), effect size index (ESI), standard response mean (SRM), and the relative efficiency index (REI). The relation between the AUC of the ECLAM and the SLICC was investigated by linear modelling.

Results—The ECLAM and SLEDAI are responsive to changes in DA irrespective of the type of responsiveness statistic used (ES all >0.96, ESI all >0.9, and SRM all >0.5). The ECLAM is more sensitive than the SLEDAI (REI all >1.99; α value (D_{oc}/F) = 0.33; α value (D_{6m}/F) <0.0001; α value (D_{oc}/F) <0.0001). Like the SLEDAI, the AUC of the ECLAM is a highly significant predictor of damage in cSLE ($R^2 = 0.25$, $p < 0.0001$).

Conclusions—The ECLAM and the SLEDAI are both highly sensitive to clinical change. The ECLAM may be more responsive to change in DA than the SLEDAI. However, the SLEDAI may be a better predictor of damage in cSLE and is easier to use.

4.11 Accumulated disease activity and steroids are risk factors for damage in childhood onset systemic lupus erythematosus (cSLE)

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Background—The disease activity (DA) measured by the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) estimates the degree of inflammation in cSLE. Patients with cSLE can develop permanent disease damage. Damage in adult SLE is measured by the Systemic Lupus International Collaboration Clinics/American College of Rheumatology Damage Index (SLICC). Currently, it is unclear how the observed DA relates to the associated damage in cSLE and whether there are other risk factors for damage.

Objectives—(a) To show that accumulated DA, as measured by the area under the curve of the SLEDAI scores (AUCS) is related over time to the observed damage scored by the SLICC. (b) To perform an initial validation of the SLICC for use in cSLE. (c) To identify other risk factors for damage.

Methods—Continuing DA and damage of an inception cohort of 66 patients with newly diagnosed cSLE was measured (diagnosis between 1992 and 1999). The relation between the AUCS and the SLICC was investigated by simple linear modelling and multivariable models were generated to identify other risk factors for damage, like age at diagnosis, race, sex, drugs, duration of disease, hypertension, antiphospholipid antibodies (aPL) and kidney disease. A survey of physicians working in the cSLE clinic was made to validate the SLICC when used in children.

Results—13/14 physicians found the SLICC suitable for use in cSLE. The AUCS is a highly significant predictor of damage ($R^2 = 30\%$, $p < 0.0001$). When multivariable modelling was used ($R^2 = 50\text{--}57\%$) it was shown that steroids contribute significantly to damage (standard regression β coefficient (STB) = 0.44–0.61; $p < 0.0001$) as may aPL (STB = 0.18–0.20; $p < 0.02$). Second line agents may protect from damage

(–STB = 0.22–0.23; $p < 0.0001$). Race, sex, hypertension, duration of disease, and age at diagnosis were not important risk factors.

Conclusions—The SLICC has face, content, and construct validity for cSLE. Continuing DA in cSLE results in damage. Steroids significantly increase and second line agents decrease damage.

4.12 Preliminary criteria for defining disease flare in patients with juvenile rheumatoid arthritis (JRA)

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Background—The proportion of patients with disease flare and time to disease flare have been used as primary efficacy measures in randomised controlled trials of new therapeutic agents for JIA. The definitions of disease flare, however, have been developed using limited datasets, and none has undergone prospective validation.

Objective—To develop preliminary criteria for defining disease flare in patients with polyarticular JRA.

Methods—We used data from a 4 month, placebo controlled, randomised withdrawal trial of etanercept in patients with a polyarticular course of JRA ($n = 51$). The JRA core set of response variables was used to derive flare criteria—that is, number of joints with active arthritis, number of joints with limited range of motion, Childhood Health Assessment Questionnaire (CHAQ), erythrocyte sedimentation rate (ESR), doctor's global assessment of overall disease severity, and parent/child assessment of overall wellbeing. The treatment with placebo was used as criterion standard of flare. Receiver operator curves, positive and negative predictive values (PPV and NPV), and simple generalised estimation equations (GEE) were used to identify and quantitatively assess the validity of various disease flare criteria.

Results—Worsening in 3 of 6 core set variables by $\geq 30\%$ without improvement in more than 1 of the remaining variables by $\geq 30\%$ (the definition of flare used in the etanercept study, referred to as definition I) had a sensitivity of 77% and specificity of 76% (PPV = 0.77; NPV = 0.76). The highest sensitivity (88%) was obtained by defining disease flare as $\geq 50\%$ worsening in any 2 core set variables (referred to as definition II). By GEE we identified definition III (any worsening in the CHAQ, worsening of ESR by $\geq 30\%$ and worsening in the active joint count by $\geq 10\%$) as a being among the most valid (sensitivity = 62%; specificity = 92%; PPV = 0.85; NPV = 0.71). Although we identified other definitions with acceptable properties, definitions I–III seemed to be the most useful.

Conclusions—We propose definitions I–III as the most suitable and suggest that these criteria of disease flare undergo further confirmation through prospective studies.

4.13 Accuracy of functional outcome measures in defining improvement in juvenile idiopathic arthritis

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Recently, emphasis has been placed on the need to include a functional measure in clinical trials of patients with JIA. In recent trials the childhood health assessment question-

naire (CHAQ) and the child health questionnaire (CHQ) have been included. We studied patients with JIA followed prospectively in our clinic to determine the ability of these 2 measures and that of the juvenile arthritis quality of life questionnaire (JAQQ) to define improvement before and after a treatment of expected efficacy.

Fifty patients with JIA were included and were entered at start of new drug treatment. Six standard measures were administered simultaneously—the active joint count, sum of joint severity score, pain (5 point scale), patient/parent global assessment (5 point scale), physician global assessment (5 point scale), and the erythrocyte sedimentation rate—in addition to the CHAQ, CHQ, and the JAQQ, at 6-weekly intervals up to 18 weeks. Patients were defined as improved or not improved at each time point based on a composite of the 6 standard measures (gold standard). Improvement in each of the CHAQ, CHQ, and JAQQ was defined as an improvement from baseline of at least 20%.

Level of agreement between each of the functional measures and the gold standard was assessed by the κ statistic. κ Values for each of these 3 measures at 6, 12, and 18 weeks, respectively, were as follows: CHAQ (0.03, 0.40, and 0.36), CHQ (0.42, 0.50, and 0.45), and JAQQ (0.53, 0.50, and 0.53). These data suggest that the JAQQ may be more accurate than the other 2 measures and thus might merit inclusion in clinical trials.

4.14 Growth and final heights in patients with systemic forms of juvenile chronic arthritis, given long term steroid treatment

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Objective—To evaluate the spontaneous growth and the final heights of patients with systemic forms of juvenile chronic arthritis (JCA) and receiving steroid treatment from the onset of the disease.

Patients and methods—Heights were expressed as SDs for chronological age (CA), and were assessed yearly during the disease, after discontinuing steroid treatment and until final heights. Final heights were compared with target heights. 24 patients were studied: 15 girls, 9 boys. Their mean (SD) CA at diagnosis was 3.4 (2.4) years, the steroid treatment was started at a mean (SD) CA of 3.5 (2.3) years, the mean duration of the steroid treatment and of the follow up were respectively 6.5 (3.6) years and 13.7 (5.1) years. The mean (SD) prednisone dose decreased from 1 (0.5) mg/kg/d in the first year to 0.3 (0.3) mg/kg/d in the fourth year of the disease.

Results—At the onset of the disease, mean (SD) height was -0.03 (1.3) SD and did not differ significantly from mean target height (-0.3 (0.1) SD). During the disease, there was a significant loss of height from -0.03 (1.3) SD at the onset of the disease to -2.4 (1.2) SD ($p<0.001$) after 4 years of follow up. The mean loss of height during the whole period of steroid treatment was -2.7 (1.5) SD and correlated with the duration of treatment ($p<0.01$, $r^2=0.2$). After discontinuing steroid treatment, 70% of the patients (17/24) experienced a partial catch-up growth with a mean gain of height of 1 (0.5) SD, but 30% (7/24) had a persistent loss of height of -0.7 (0.4) SD. The mean final height of the cohort

was -2.1 (1.9) SD with a mean difference between final height and target height of -1.7 (1.6) SD. Mean final height correlated with mean height at the end of steroid treatment ($p<0.001$).

Conclusions—(a) a significant loss of height was seen in patients with systemic forms of JCA; (b) a partial catch-up growth can occur after recovery, but final heights of the patients remained far from their target heights.

4.15 Ethnic differences in the incidence of Kawasaki disease, Henoch-Schönlein purpura, and other vasculitis in 590 children

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Objective—To determine the incidence and ethnic distribution of Henoch-Schönlein purpura (HSP), Kawasaki disease (KD), and rare vasculitides in children resident in one region of the UK.

Methods—Monthly questionnaires were sent prospectively to 321 consultants over 3 years (return rate 71.1%). Case ascertainment was verified by review of a further 393 case notes with diagnostic codes for vasculitis and a single questionnaire to 2860 primary care doctors (GPs) (return rate 59.7%).

Results—590 new cases of vasculitis fulfilling established diagnostic criteria were collected prospectively from the 1.2 million children in the region. Asian children had a higher incidence of KD, systemic lupus erythematosus (SLE), and primary systemic vasculitis (PSV) than white children (table 2).

The annual incidence of HSP was higher than previously described, rising to 22.1/100 000 under 14 years (95% CI 19.2 to 25.0) and 70.3*/100 000 between the ages of 4 and 6 years (95% CI 54.2 to 86.4). The annual incidence of KD rose to 5.5/100 000 in children under 5 (95% CI 3.1 to 7.8), and 14.6*/100 000 (95% CI 1.3 to 27) in Asian children under 5 years. In girls over 11 years the annual incidence of SLE was 12.3*/100 000 (95% CI 7.3 to 17.3).

Conclusion—Our study has recognised a higher incidence of vasculitis than previously reported in children, particularly in children of Asian origin.

4.16 Outcome measures in adolescents attending a unique residential programme for adolescents with diffuse and localised idiopathic pain syndromes (fibromyalgia and reflex sympathetic dystrophy)

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Introduction—Diffuse and localised idiopathic pain syndromes (fibromyalgia and reflex sympathetic dystrophy) account for a significant amount of morbidity in adolescents and their families. We have developed a unique

multidisciplinary residential programme that focuses on physical and cognitive aspects of rehabilitation in these complex conditions. Here we present outcome data taken before the programme and at 3 and 6 months after the programme.

Methods—Over a period of 12 months 20 adolescents with chronic pain syndromes that had caused significant morbidity for at least 4 years were referred to one of our 3-week residential programmes. Each young adult at the pre-programme, 3 month, and 6 month post-programme assessments completed visual analogue pain scales, functional disability inventory questionnaires, and Spence Children's Anxiety Scale questionnaires. School attendance data before and after the programme were also evaluated.

Results—The pre- and post-programme data were analysed using Wilcoxon signed ranks tests in the 20 adolescents and 20 accompanying adults. The children's anxiety scales showed a significant improvement in social phobia ($p=0.03$), separation anxiety ($p=0.007$), and physical injury fear ($p=0.04$) after the programme. Analysis of the functional disability inventory data of both the young adult and accompanying adult scores showed significant improvement after the programme ($p=0.05$, $p=0.02$ respectively). There was no significant improvement in pain using the visual analogue scales ($p=0.32$).

Discussion—Our outcome data show that, with a structured cognitive and physical therapy residential programme, adolescents can develop strategies allowing them to reduce their disability and participate increasingly in daily activities. Parental and adolescent anxiety decreased and school attendance improved significantly in all cases; this was despite the pain scores remaining unchanged at 6 months after the programme.

4.17 Evaluation of the Norwegian version of the Child Health Questionnaire (CHQ) in children with juvenile arthritis

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Objective—To assess the reliability and validity of the Norwegian version of the generic Child Health Questionnaire-parent form (CHQ-pf50) in patients with early juvenile arthritis.

Methods—Parents of 92 children remitted to The National Hospital, Oslo (median age 8.5 years, range 4–16) with juvenile arthritis (JA) and a mean disease duration of 11 months (range 1–29) and 97 parents of healthy controls, completed the Norwegian version of the CHQ-pf50 at baseline. The patients were reassessed after a mean of 10 months (range 3–20).

Results—The internal consistency of the CHQ-pf50 was good (Cronbach's $\alpha=0.84$). Parent-patient and test-retest correlations of the underscores were moderate to high

Table 2 Annual incidence of vasculitis (95% CI) per 100 000

Ethnicity (% region)	HSP (n=508)	KD (n=73)	SLE (n=28)	PSV (n=8)
All	20.4 (17.8 to 23.0)	2.1 (1.3 to 2.9)	0.8 (0.3 to 1.3)	0.24 (0.05 to 0.52)
Asian (10)	24.0 (16.2 to 31.5)	4.9* (0.9 to 8.9)	2.8 (–0.24 to 5.8)	1.15* (–0.8 to 3.1)
Black (3)	6.2* (–2.4 to 14.8)	2.1 (–2.9 to 7.1)	5.2 (–2.7 to 13.1)	—
White (86)	17.8 (15.3 to 20.3)	1.9 (–0.8 to 4.6)	0.5* (0.03 to 0.87)	0.14 (–0.1 to 0.38)

* $p<0.01$.

(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