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

the adductors, vasti and rectus femoris muscles. In several cases there was swelling of the affected muscles. These muscles were shown to be hyperaemic by colour Doppler imaging. After initial steroid treatment all USS showed a remarkable reduction in muscle echogenicity, muscle swelling, and vascularity. Two USS showed marked wasting and fatty infiltration after treatment and a major increase in subcutaneous adipose tissue. In one patient a clear progression from echogenic hyperaemic muscle through reduced echogenicity to fatty atrophy was seen over serial scans. One patient had calcinosis on follow up. The USS findings correlated well with MRI and biochemical markers of disease activity.

Conclusion—Musculoskeletal ultrasound is a safe, inexpensive, non-invasive means of supporting the clinical diagnosis of JDM and of monitoring disease progression, without needing sedation, which is often needed for an MRI in young children. Ultrasound findings appear to correlate well with other markers of disease activity.

6.2 FK-506 in the treatment of unresponsive juvenile dermatomyositis

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Objective—To describe the efficacy and safety of FK-506 (tacrolimus) in three children with dermatomyositis.

Patients and methods—Patient 1: 7 y 8 m old girl, with no resolution of the skin abnormalities, strength 4/5 after 2.33 years of disease, previously treated with high dose steroids, methotrexate and cyclosporin A (CyA).

Patient 2: 11 y 7 m old girl, with impressive lack of strength (2/5) after 1.16 years of treatment (high dose steroids and CyA); hypertension as CyA side effect.

Patient 3: 11 y 11 m old boy, longstanding disease (7.83 years), muscle strength 4/5 and persistence of severe skin lesions

In all three FK-506 was started at 0.1 mg/kg/day and levels (5–10 ng/ml) were monitored every three months. Parent's consent was mandatory to start treatment.

Results—All three patients responded extremely well to FK-506. Muscle strength was considered normal (5/5) for all three after one year of treatment. Skin lesions disappeared in patients 1 and 2. In patient 3, residual atrophic skin areas are still visible. Patients 1 and 2 have stopped steroid treatment, while patient 3 went from 10 mg/day to 5 mg/48 h. In the only patient with raised lactate dehydrogenase at the beginning of treatment (patient 2), levels normalised after 6 months. Neither major nor minor side effects occurred
Conclusion—FK-506 might be a good alternative for patients with dermatomyositis who do not respond to or are intolerant of high dose steroids or CyA.

6.3 Familial occurrence of juvenile dermatomyositis

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A case of familial dermatomyositis (DM) is described, in which all affected family members had onset of symptoms during infancy. Father and daughter have mild dermatologi-

cal stigmata of DM, with no muscle involvement. The son has juvenile dermatomyositis with biochemical and histological evidence of myositis at diagnosis and florid cutaneous manifestations. No HLA or DQ associations were noted in this family. Familial DM is rare, early onset of disease in infancy in all affected family members has not previously been described.

6.4 Risk factors at onset in juvenile dermatomyositis: correlation with prognosis

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We describe 33 patients (18 female, 15 male) affected with dermatomyositis, with a mean age at onset 7 y 1 m (range 2 y 3 m–17 y 5 m) and mean period before the diagnosis 4 m (range 7 d–2 y 1 m).

The evaluation of both signs and symptoms and the muscle enzymes values at the onset allows a subdivision into 2 groups: 16 patients had an acute onset (skin rash, high fever, remarkable muscle weakness, nasal speech, dysphagia, skin vasculitis, respiratory failure, and large increase of skeletal muscle enzymes in serum), and 17 patients had an insidious onset (skin rash, arthralgia, less marked muscle weakness). Six out of 16 patients with acute onset treated with pulses of methylprednisolone, plasmapheresis, IV immunoglobulin, and different immunosuppressive treatment (methotrexate, cyclosporin, cyclophosphamide), also required intensive care treatment, and 1 required surgery for intestinal vasculitis. Three out of six patients who required intensive care treatment died, two for respiratory failure and one for systemic vasculitis.

In contrast, all the patients with an insidious onset improved with methylprednisolone alone.

In conclusion, early and aggressive treatment is mandatory in the high risk subtype to obtain a more rapid clinical and enzymatic improvement and a shorter course.

6.5 Juvenile dermatomyositis (JDM) in 23 patients

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We retrospectively studied 17 girls and 9 boys with JDM followed up at the paediatric rheumatology unit since 1982. Mean (SD) age at disease onset was 5.8 (2.9) years (range 1–11.8), at diagnosis 6.4 (2.7) years (2.4–12). Disease duration at diagnosis was 7 (8) months (0–27). All patients but one had typical skin lesions, 18 had proximal muscle weakness. Other clinical symptoms at diagnosis were fever and weight loss (14), muscle pain (10), arthralgia/arthritis (9), dysphagia (3), calcinosis (2), tendon retraction (1), tachycardia (1), hepatomegaly (1). All patients but one were treated with oral prednisone. Other drugs were methylprednisolone pulses (11), cyclosporin A (6), methotrexate (6), hydroxychloroquine (5), IV immunoglobulin (5), non-steroidal anti-inflammatory drugs (2). Nineteen patients responded well to the initial treatment. A mean of 2.6 relapses (range 1–5) occurred in 15 patients either during treatment (30 relapses) or after treatment was discontinued (9 relapses).

Clinical symptoms seen during follow up were persisting skin lesions (26), muscle weakness (21), muscle pain (14), arthralgia/arthritis (12), calcinosis (12), dysphagia (8), fever or weight loss (6), tendon retraction (7), cutaneous staphylococcus infection (7), clinical lipodystrophy (2), and pancreatitis (1). At the last visit 8 patients had stopped treatment without sequelae, 6 had stopped treatment with sequelae, 12 were still receiving treatment after a mean disease course duration of 3.5, 7.9, and 4.3 years respectively.

6.6 Treatment of juvenile dermatomyositis (JDM) with high dose oral steroids or with steroid pulses and low dose oral steroids

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Objective—Treatment with oral steroids may control inflammation in the large majority of children with JDM, but is followed by Cushingoid syndrome.

Methods—Prospective randomised open study of oral high dose steroids (prednisone 2 mg/kg for 4 weeks followed by gradual decrease) versus repeated pulses (IV methylprednisolone 20 mg/kg for 3 days with decreasing frequency of pulses) plus low dose oral steroids (prednisone 0.2 mg/kg). Patients were evaluated after 8 weeks for initial response and for a further 40 weeks for relapse.

Results—24 patients (18 girls) were enrolled, median age 7 years, range 3–15. 13 patients received steroid pulse treatment, 11 high dose oral steroids. All patients were considered responders. 19/24 patients were followed up for ≥48 weeks, the remaining patients for 8–38 weeks (3 × pulse, 2 × oral). 7/24 patients had a relapse (3 × pulse, 4 × oral). Cushingoid syndrome was found in 9/11 patients receiving oral steroids and in 3/13 receiving pulse steroids.

Conclusion—Treatment of JDM with pulse steroids plus low dose oral steroids may be as effective as high dose oral steroids, but the frequency of steroid adverse effects may be diminished.

Participants—Biedermann/Berlin, Eisenberg/Herford, Häfner/Garmisch, Häfner/München, Haller/Schweiz, Heidemann/Augsburg, Horneff/Halle, Keitzer/Berlin, Lehmann/Bad Bramstedt, Leipold/Erlangen, Leupold/Dresden, Möbius/Cottbus, Queisser/Ludwigshafen, Quetzsch/Plauen, Weißbarth-Riedel/Hamburg.

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6.7 Low dose short term corticosteroid treatment for "amyopathic" childhood dermatomyositis (DM): the role of MRI

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In 1975 Boham and Peter proposed rash and 1 to 3 out of 4 possible myositis-defining criteria (weakness, increased muscle enzyme, positive biopsy, EMG) for classification of dermatomyositis (DM). In the largest series published to date by LM Pachman *et al* all children depicted at least one myositis-

defining criterion and all presented at least one abnormal muscle enzyme.¹ Classically, patients who do not fulfil the myositis-defining criteria are called amyopathic.

In this report we present 4 children who presented with typical DM rash, normal muscle enzymes, and no history of weakness. The presence on magnetic resonance imaging (MRI) of a typical abnormal signal intensity on fat suppressed fat spin echo T2 weighted image circumvented the need for invasive studies (EMG/biopsy) by rendering us confident of the diagnosis. We treated these children with a short course of prednisone (3 months total) at a low dose (0.5–1 mg/kg for 4–6 weeks followed by tapering) because of our conviction that they had mild disease. Follow up MRI showed resolution at 1–2 months of treatment and follow up at 6–24 months from onset disclosed no recurrence and no calcinosis. All patients are receiving hydroxychloroquine and none is receiving corticosteroids. Toxicity was minimum.

Conclusions—1 MRI should be considered a candidate non-invasive diagnostic criterion for DM; 2 perhaps, milder disease may be treated less aggressively than classic DM.

1 Pachman LM, *et al.* J Rheumatol 1998;25:1198–204.

7 Juvenile idiopathic arthritis

7.1 Changes of antioxidative status and free radical damage in subsets of juvenile idiopathic arthritis (JIA) over a period of three months

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JIA is an inflammatory disease in which the role of free radicals has not yet been clarified. We recently showed a specific pattern of antioxidative status and free radical damage in subsets of JIA.¹ We investigated whether this pattern is characteristic for subsets of disease or inflammatory activity.

Patients (n=85) with JIA were seen consecutively in the paediatric rheumatology unit during one year and again after three months. The disease subsets were: oligoarticular juvenile rheumatoid arthritis (oJRA, n=54), polyarticular JRA (pJRA, n=3), systemic JRA (sJRA, n=4), spondyloarthritis (SA, n=18), and psoriatic arthritis (n=6). Patients with non-inflammatory joint pain served as controls (n=15). Erythrocyte sedimentation rate (ESR), in vitro radical resistance of erythrocytes (RRE), total radical trapping ability of plasma (TRAP), malondialdehyde as marker of lipid peroxidation, sulphhydryl (SH) groups, and α -tocopherol as antioxidants were analysed in blood.

According to the JIA subsets, the following differences were seen between the first and second visit: a decrease in the tender joint score in patients with oJRA ($p<0.001$) corresponding with an increase of α -tocopherol ($p<0.05$) and TRAP ($p<0.01$). All patients with JIA were stratified according to inflammatory disease activity into groups with low (<20 mm/1st h), medium (20–40 mm/1st h), and high ESR (>40 mm/1st h). The differences seen between these groups and controls during the first visit (increase of in vitro RRE (high ESR); and decrease in SH groups (medium ESR)) were not seen again during the second visit.

The increase in antioxidative potential and simultaneous decrease of tender joint score in patients with oJRA underline the role of antioxidants in JIA. It remains to be elucidated whether the change of pattern according to inflammatory disease activity means that there is no characteristic reproducible pattern of antioxidative status and free radical damage in relation to inflammatory disease activity.

1 Arthritis Rheum 1999;42(suppl):S181.

7.2 Growth disturbances in patients with juvenile idiopathic arthritis (JIA): Has the prevalence changed?

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Previous studies have shown that a significant proportion of patients with juvenile idiopathic arthritis (JIA) develop growth disturbances. In recent years the treatment of JIA has become more aggressive, mainly through the increased use of methotrexate (MTX). Therefore, we recently examined the growth parameters of 103 consecutive outpatients with JIA. We correlated the height and weight centiles with demographic data, disease characteristics, and drug use. The patients' mean (SD) age was 12.3 (6.2) years, disease duration 6.8 (5.2) years, height 142.6 (27.7) cm, and weight 45.0 (24.9) kg. The mean height centile for all patients was 45.7 (31.5) and for weight it was 53.4 (30.8). Overall, 16% of the patients were 5th height centile ($p=0.03$ compared with the normal population). Height centiles were significantly less in patients with systemic JIA ($p<0.001$) and also correlated with steroid use ($p<0.001$). Weight was significantly correlated only with disease subtype ($p=0.003$). Neither height nor weight centiles correlated with MTX use (48% of the patients used MTX). Despite more aggressive treatment a significant proportion of our patients had growth, mainly height, disturbances. However, the mean height and weight centile of the entire JIA cohort was nearly normal.

7.3 Study of IL6, TNF, and IFN γ in the serum of patients with juvenile idiopathic arthritis and systemic lupus erythematosus

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We evaluated the levels of interleukin 6 (IL6), tumour necrosis factor (TNF) and interferon γ (IFN γ) in 41 patients with juvenile idiopathic arthritis (JIA)—13 with polyarticular, 15 with pauciarticular, and 13 with systemic onset type. The serum concentration of TNF was determined in 21 children with systemic lupus erythematosus (SLE).

We found raised serum IL6, TNF, and IFN γ levels in all onset types of active disease. The highest concentration of IL6 was established in systemic JIA as compared with polyarticular and pauciarticular JIA ($p<0.001$). The level of IL6 was significantly higher in active disease than in inactive disease ($p<0.001$) and correlated with systemic inflammatory activity. A significant correlation between IL6, C reactive protein, and erythrocyte sedimentation rate in serum was found ($p<0.001$).

Our results show that the serum level of IL6 may be a marker of active disease and have a role in the regulatory pathway of inflammation in JIA.

We concluded that the disease activity of SLE and the absence of previous long term immunosuppressive treatment are associated with increasing levels of TNF.

Results from the analysis of cytokine expression may provide a basis for the use of specific anticytokine treatment.

7.4 Referral of children with JRA by general paediatricians

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Background—There is limited information known about the role of general paediatricians in the care of patients with JRA.

Objectives—To explore paediatricians' self reported treatment and referral patterns for patients with juvenile rheumatoid arthritis (JRA) and to identify factors associated with the referral of patients for all JRA care.

Methods—Self administered surveys were mailed to a national random sample of 700 paediatricians. Subspecialists and those who do not see patients were excluded. The 4 page survey included demographic information, questions about JRA educational and clinical experience, and factors influencing referrals. Response rate was 50%.

Results and conclusions—General paediatricians refer patients with JRA for diagnosis and treatment at a high rate, even though many rate themselves as comfortable in diagnosing JRA. Most paediatricians (90%) refer patients with JRA to paediatric rheumatologists. Factors cited as important in patient referral were identified. Residency training experience with patients with JRA did not decrease the likelihood of referring patients for all JRA care. Most paediatricians reported the need for a practice guideline for the primary care management of JRA. These data suggest that most general paediatricians feel they have a limited role in the care of children with JRA.

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7.5 Infection associated MAS in 3 patients receiving ASCT for refractory JIA

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Autologous stem cell transplantation (ASCT) has been proposed as a possible treatment for severe autoimmune diseases. In 1997 we started a study on the efficacy of ASCT on disease activity in children with refractory polyarticular or systemic juvenile idiopathic arthritis (JIA). So far 10 children with systemic JIA and 2 with polyarticular JIA, all with progressive disease activity despite the use of corticosteroids—methotrexate up to 1 mg/kg/week and cyclosporin A (2.5 mg/kg/day) were treated with ASCT with a follow up of 1–30 months.