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 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.

Methods—In this first part of a prospective study to obtain data about HLA class I and II in patients with the different major manifestations of ARF, 117 patients with SC were tested for HLA class II antigens by the polymeric 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 χ² 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 (χ² with Mantel-Haenszel = 7.61; p < 0.006).

Conclusion—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 arthritis was detected and polyarthritis 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 aortal valves in 10 (9%). In 5 (5%) cases pancycarditis 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.

Conclusion—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/aortal valves) and in 5 patients with ReA; one patient developed juvenile rheumatoid arthritis (pauci-type II) and in one case juvenile anklyosing spondylitis was diagnosed. The mean age at diagnosis was 14.3 years and 58 (54%) were male, 5 (5%) were female. 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 arthritis was detected and polyarthritis 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 aortal valves in 10 (9%). In 5 (5%) cases pancyarditis 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.

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6.2 FK-506 in the treatment of unresponsive juvenile dermatomyositis

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We describe 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 abnormality after 2.3 years of disease, previously treated with high dose steroids, methotrexate and cyclosporin (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.

All 3 patients with FK-506 were started at 0.1 mg/kg/day and levels (5–10 ng/ml) were monitored every 3 months. Patient’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 dermatological 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 dermatomyositis 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, etanercept, infliximab) 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 6 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 at onset with 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 3.9, 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 of 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.


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6.7 Low dose short term corticosteroid treatment for “amypathic” childhood dermatomyositis (DM): the role of MRI

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In 7 Bohan 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 myosit-
The increase in antioxidative potential and simultaneous decrease of tender joint score in patients with oJRA underline the role of anti-oxidants 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.

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

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