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 (317.0 (118.4) mIU/ml) There was no difference between the sexes in vaccine responsiveness. When antibody levels between two different 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 mechlorexate or prednisolone treatment. However, there was a negative correlation between prednisolone dose and anti-HBs titre ($r=-0.23$).

Conclusions—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 choræa

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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 choræa (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 specific primers technique, with genomic polymerase chain reaction with sequence-specific amplification allowing the diagnosis of acute infection using a single serum sample, confirmed by a parallel serum in 4/7 patients. Immuno- globulin 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 58% of patients with SC. 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 onset spondylodiscitis was diagnosed.

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 onset spondylodiscitis was diagnosed. Five (6%) cases mononuclear cells were 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 occurs with endocarditis (damage of the mitral or aortal valves) and had 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 streptococcal infection) is a disorder usually presenting as neuropsychiatric symptoms resulting from autoimmune responses to streptococcal (group A $\beta$ 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 $\beta$ 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 and subsequently developed emotional lability, cognitive deficits, oppositional behaviours, and motor hyperactivity. Investigation showed mitral valve stenosis, 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.

Conclusions—The PANDAS case corresponds with described diagnostic criteria (Kleiner- ser, 1999, Schuman, 1999).

6 Juvenile dermatomyositis

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

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Objective—To assess the value of ultrasound scans (US) 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—In 7 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 findings. USS were correlated with MRI of the thigh in all patients.

Results—All 5 USS performed at diagnosis were abnormal. Most showed diffuse symmetric 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 M CUSTODIO*, M BORONAT†, S MARCEL*, C ARNAL GUILERA, C MODESTO*, M BORONAT†, M BORONAT†

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 vascularity, 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, 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 but 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 3.9, 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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Support for the study was provided by the Deutsche Kinder- und Jugendforschungsgemeinschaft.

Methods—Prospective randomised open study of oral high dose steroids (prednisone 2 mg/kg for 4 weeks followed by gradual deceleration) 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 “amyopathic” childhood dermatomyositis (DM): the role of MRI

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In 13 children, 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 antioxidant 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
anti oxidant 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?

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Previous studies have shown that a significant proportion of patients with juvenile idio-
patic arthritis (JIA) develop growth distur-
ances. 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 char-
acteristics, and treatment. The patients' mean (SD) age was 12.3 (6.2) years, disease dura-
tion 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 (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 propor-
tion 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 IL-6, TNF, and IFN-γ in the serum of patients with juvenile idiopathic arthritis and systemic lupus
erthematous

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We evaluated the levels of interleukin 6 (IL-6),
tumour necrosis factor (TNF) and interferon
γ (IFNγ) in 41 patients with juvenile idiopathic arthritis (JIA)—13 with poly-
articular, 15 with pauciarthritic, and 13 with
systemic onset type. The serum concentra-
tion of TNF was determined in 21 children with
systemic lupus erythematosus (SLE).
We found raised serum IL-6, TNF, and
IFNγ levels in all onset types of active disease.
The highest concentration of IL6 was estab-
lished in systemic JIA as compared with poly-
articular and pauciarthritica (p<0.001).
The level of IL6 was significantly higher in active disease than in quiescent disease
(p<0.001) and correlated with systemic inflammatory activity. A significant correla-
tion 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 treatments 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 paediatrici-
cians 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 paediatrici-
cians refer patients with JRA for diagnosis and
treatment at a high rate, even though
many rate themselves as comfortable in diag-
nosing JRA. Most paediatricians (90%) refer
patients with JRA to paediatric rheumatolo-
gists. 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 paediatrici-
cians feel they have a limited role in the care
of children with JRA.
This work was supported by a grant from the
Arthritis Foundation.

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—metho-
trexate 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.