ups, he was well. Three months later, a widespread enlargement of mesenteric lymph nodes and a thickening of colonic walls were shown. In the following months we noted (a) high IgA plasma concentration (9.45 g/l); (b) increased mevalonate urinary excretion; (c) strongly reduced activity of mevalonate kinase (5.4 v 347 pmol/min/mg). Familial Mediterranean fever was ruled out by genetic analysis. On this basis, we suggested the diagnosis of hyper-IgD syndrome. The patient was treated with colchicine (1 mg/day continuously), prednisone (0.5 mg/kg), and naproxene (15 mg/kg) only at the beginning of flare up. Table 1 shows the results obtained.

Table 1

	Flare ups (days) Mean (SD)	Follow up (months)	Flare ups
No treatment	17 (8.2)	5	7
Colchicine	33 (25)	15	15
Prednisone	14 (6.2)	5	9
Naproxene	18 (7)	3	3

Although our one case does not permit statistical analysis, our data seem to suggest that colchicine gives better disease control, reducing fever flare ups, whereas prednisone and naproxene sharply stop the fever attack at the beginning.

We thank RA Wanders for the mevalonate-kinase assay.

9.19 Joint involvement in eosinophilic gastroenteropathy in childhood

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Eosinophilic gastroenteropathy (GE) is an uncommon disease characterised by eosinophilic infiltration of the gut wall. The disease may have different clinical presentations. Joint involvement is likely an underestimated complication of GE.

We report on 13 patients seen during the period 1992–99. Their mean age at the presentation of the disease was 8 months (range 1–4). All patients underwent gut endoscopy with multiple biopsies. We considered the gold standard to be villous containing >20 eosinophils infiltrating the epithelium.

The heralding symptoms were severe iron deficiency anaemia in 5 patients, which was associated with oedema due to loss of protein through diarrhoea in a further 5 patients; and haematochezia in 5 patients. Two patients presented severe bloody diarrhoea. The remaining patient came to us because of exudative ascites.

Four patients developed non-erosive arthritis in both the knee (2) and at the tibiotarsal joints. This symptom occurred after 12–19 months from the diagnosis. Arthritis was treated with sodium naproxene in 2 patients and intra-articular steroid infiltration in 1 patient. Interestingly, 1 patient developed a good response to an exclusive monomeric diet; when this schedule was modified arthritis flared up. No patient needed steroids or immunosuppressive drugs for the control of arthritis.

Our experience suggests that (a) arthritis is a relatively common complication of GE; (b) the feeding treatment using monomeric dietary schedule may be effective in the treatment of GE related arthritis.

10 Scleroderma

10.1 Thermography in juvenile localised scleroderma assessment

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Aim—To evaluate the clinical use of infrared thermography in localised scleroderma (LS) in disease activity assessment and management.

Methods—We retrospectively reviewed thermal images of children with LS obtained between 1993–2000. Thermographs were included only when a contemporary detailed clinical description of the lesion(s) was available. Lesions were classified as “active” (new or extended) or “quiescent” according to clinical description (colour, skin texture, measurements). Thermographs were considered positive when the affected area temperature was >0.5°C higher than the surrounding skin or the opposite site. Two clinicians (GM and KJM), blinded to the clinical description and thermography report, reviewed the thermal images independently. Full agreement in scoring was achieved in 86%, and discordant results were resolved by mutual examination.

Results—40 patients were included in the study (26 F, 14 M). The most common diagnosis was a combination of morphea and linear scleroderma (M+LiS, 14 patients), followed by isolated LiS (11 patients), en coup de sabre (6 patients), and M (5 patients). 68 lesions were examined: 33 affecting the legs, 16 the arms, 10 the face/scalp, and 9 the trunk. We reviewed 130 separate thermal images, 34 lesions having multiple thermographic examinations.

There was complete agreement between the clinical description and thermography in all new lesions (table 1). Of the clinically inactive lesions positive on thermography, most were “old” lesions with the presence of severe atrophy and subcutaneous fat loss.

Table 1

	Thermography	
	Positive	Negative
Clinically active	Pos 47/51 (92%)	4/51 (8%)
Inactive	Neg 25/79 (32%)	54/79 (68%)

Conclusions—Infrared thermography is a potentially reliable tool for assessing the activity of LS lesions in conjunction with clinical activity, particularly for clinically suspicious new and extending lesions. Further evaluation is needed to determine whether thermography can predict future progression of lesions, particularly those which are equivocal clinically.

10.2 An unusual type of scleroderma with neurological disease

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We describe the case of a 14 year old boy who started with a linear scleroderma and presented later a neurological disease.

At the age of 11 years the boy developed a marked hypotrophy and hypoplasia of the right forearm apparently owing to prolonged immobilisation. Over a period of few months the disease progressed to affect the skin and muscles of the right arm, hand, and fingers with a sclerotic evolution. All laboratory and radiological examinations (x rays and magnetic resonance imaging (MRI) of the arms, x rays of the chest and of the gastrointestinal tract), spirometry, and a nailfold capillaroscopy were normal except for a high antinuclear antibody titre (1/1280).

Borrelia infection (IgM and IgG low titre) was suspected and the patient began treatment with penicillin 15 000 000 U/d for 10 days, without any improvement.

Two years later he had uveitis and seizures (abnormal EEG, cranium asymmetry at x ray examination, microvascular ischaemic encephalopathy at MRI, negative cerebral arteriography).

Recently, facial hemiatrophy with a contralateral hemisindrome is evident and skin and muscle atrophy have worsened.

Whether this form is an evolution of linear systemic scleroderma or is linear scleroderma with neurological disease, which has already been described, cannot be stated for sure.

10.3 Efficacy and safety of autologous peripheral blood stem cell transplantation in three children with systemic sclerosis and progressive pulmonary disease

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Background—Autologous peripheral blood stem cell (PBSC) transplantation has been proposed as a treatment for autoimmune diseases with fatal prognosis.

Patients—Three children with systemic sclerosis since the age of 4 (patient 1), 7 (patient 2), and 6 (patient 3) years had a severe disease course, with progressive cutaneous, articular, and pulmonary disease despite immunosuppressive treatment. Because pulmonary fibrosis, once established, is progressive, at the age of 11, 10, and 9 years, respectively, they discontinued all drugs and received an autologous PBSC transplantation.

Transplantation procedure—After obtaining approval from the local ethical committee and parents' informed consent the three children were given CY (4 g/m²) followed by G-CSF (10 mg/kg/d) to allow the collection of PBSC. The conditioning regimen before transplantation comprised CY at a dose of 50 mg/kg from day 5 to day 2 and the monoclonal antibody Campath-1g at a dose of 10 mg/d for 2 days. The post-transplantation period was uneventful in all patients.

Results—Two patients (patients 1 and 2) had a significant clinical benefit at 40 and 16 months, respectively, after transplantation, with improvement in growth rate and general wellbeing, and general skin softening. In both of them high resolution pulmonary computed tomography (HRCT) scan did not show any progression of fibrotic changes. At variance, in 1 patient (patient 3) the transplantation procedure did not affect significantly the skin

disease, and a repeated pulmonary HRCT after 12 months showed a further progression of pulmonary fibrosis.

Conclusion—Autologous PBSC transplantation associated with Campath-1g treatment was safe and well tolerated in all patients and resulted in marked and sustained clinical improvement in two of the three patients.

11 Treatment of paediatric rheumatic diseases

11.1 Cyclosporin A treatment in chronic recurrent uveitis in JIA

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Uveitis is a serious cause of morbidity in pauciarticular onset juvenile idiopathic arthritis (JIA), the prognosis of which is otherwise good. To assess the efficacy of cyclosporin A in the treatment of chronic recurrent JIA uveitis, where local steroids failed, cyclosporin A was given to 4 patients (2–14 years old). The initial dose was 3–5 mg/kg/24 h by mouth, followed by a 3 mg/kg/24 h maintenance dose after inflammatory suppression. Cyclosporin levels and side effect parameters (blood pressure, WBC, Cr, AST, ALT, UT) were estimated. Patients were receiving non-steroidal anti-inflammatory drugs and one of them was receiving methotrexate also. The rate and severity of recurrences were recorded before and after cyclosporin. Before cyclosporin there were 6 (range 4–7) recurrences/child, 1 recurrence/month, of mean duration 2.5 (0.5–4) months, Tyndall 3+/recurrence. After cyclosporin, there was 1 recurrence/child with Tyndall 1+/recurrence, which was the result of non-compliance. Inflammatory suppression was achieved in a mean of 25 days. Mean follow up time was 12 (6–12) months. Cyclosporin side effects were: hirsutism (3), gum hyperplasia (1). Cyclosporin A is an effective and safe treatment in persistent, refractory uveitis of JIA.

11.2 Exceptional therapeutic response to infliximab treatment in recalcitrant Crohn's spondylitis

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At age 8 years in December 1990 a white boy presented to us with limp and an erythrocyte sedimentation rate of 70 mm/1st h, a value that would not go below 60 mm/1st h for the next 9 years. The cause could not be initially established. A year later he developed ankle synovitis. 15 months later magnetic resonance imaging confirmed bilateral sacroiliitis. A negative HLA-B27 in the presence of sacroiliitis made us suspect Crohn's spondylitis,¹ but intestinal disease was not obvious until 1999. Although clinically he had only mild diarrhoea his colonoscopic and histological findings were classic of Crohn's disease. His systemic symptoms, sacroiliac pain, peripheral arthritis, and weight loss continued to deteriorate over the years despite sulfasalazine 2500 mg/d, prednisone 20 mg/d, methotrexate 17.5 mg/w, and naproxen 500 mg/d. In January 2000 he

received his first dose of infliximab 250 mg. The results of the intervention were astonishing. A month after the infusion he was able to attend school and to work part time, had no pain, no morning stiffness (previously up to 90 minutes), and his stamina normalised. There was no evidence of synovitis on examination, though lumbar mobility is still unchanged (Shober 12 cm). His response has been sustained and a second dose was given 2 months later (table 1).

Table 1

	12/90	7/99	10/99	4/00
Haemoglobin (g/l)	114	111	103	132
ESR (mm/1st h)	76	85	ND	11

Infliximab in this case was indicated for the skeletal and systemic but not gastrointestinal manifestations of Crohn's disease. A prospective study on the use of infliximab in paediatric ankylosing spondylitis/spondyloarthropathies is critical at this point.

1 J Rheumatol 1998;25:S1–49.

11.3 Nitrous oxide analgesia for intra-articular injection in children

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Background—Intra-articular injection of corticosteroid is commonly used in the treatment of inflammatory arthritis in children. Often the procedure is carried out under general anaesthetic or using sedation with agents such as benzodiazepines. Entonox, a mixture of 50% nitrous oxide and 50% oxygen, is widely used in paediatric practice for the alleviation of procedural pain. We report our experience with Entonox for intra-articular injection.

Results—Intra-articular injection was carried out in 70 patients. 69 of the patients had juvenile idiopathic arthritis and one had juvenile dermatomyositis. 64 patients (91%) tolerated the procedure well without any distress and 82 joints were injected, with a maximum of 2 joints injected each session. The joints injected were knees 73 (89%), ankles 3, wrist 1, elbow 1, metacarpophalangeal joint 1, and proximal interphalangeal joint 3. All patients who had had previous injections under general anaesthetic or with midazolam indicated that they would prefer Entonox in the future. Patients who tolerated the procedure had an age range 4 years 8 months to 17 years 7 months. There were no complications in any patient and all were fit for discharge within 30 minutes of the procedure. In 6 (9%) of the patients the procedure was not well tolerated, and 3 of these children completed the procedure in the same session using intravenous sedation. 3 of the children who did not tolerate the intra-articular injection using Entonox were aged less than 6 years.

Conclusion—In selected patients Entonox is a safe analgesic agent that may facilitate rapid and well tolerated intra-articular injection to treat inflammatory arthritis in children.

11.4 Infliximab in the treatment of persistently active refractory juvenile idiopathic (chronic) arthritis

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An open prospective trial was carried out in a young population affected by severe, refractory, polyarthritic juvenile idiopathic arthritis (JIA) to evaluate the efficacy and safety of infliximab (Remicade). We enrolled 10 female subjects, mean age 21.9 years (range 11.3–32.5), mean onset age 3.3 years (1.1–10), mean disease duration 18.6 years (9.8–31.4). All patients had been previously treated with more than one disease modifying antirheumatic drug (DMARD) (mean number 3.8/patient, as mono or combined treatment). All patients still had active disease: number of active joints (mean 5.9), erythrocyte sedimentation rate (ESR; mean 60 mm/1st h), C reactive protein (CRP; mean 39.2 mg/l), physician global evaluation (mean visual analogue scale (VAS) 52), Health Assessment Questionnaire (HAQ; mean disease index 1.43). 8 patients were still receiving corticosteroids (mean 0.1 mg/kg). All patients discontinued any other DMARD aside from subcutaneous methotrexate (mean weekly dose 12.5 mg, range 5–25) and continued with previous non-steroidal anti-inflammatory drugs and corticosteroids. Infliximab was given as a single infusion of 3 mg/kg at time 0 and at weeks 2, 6, 14, and 22. Until now 1 patient has received 5 infusions, 4 patients 4 infusions, 3 patients 3 infusions, and 2 patients 2 infusions. One patient withdrew because of a severe adverse event (hypersensitivity reaction at the third infusion). After the second infusion, 8 out of 10 patients achieved a very good response (50% reduction in number of active joints, ESR, CRP, VAS, HAQ). A significant improvement was also observed at the third infusion: number of active joints (mean 1.3), ESR (mean 21 mm/1st h), CRP (mean 1.5 mg/l), VAS (mean 13.8), HAQ (mean 1.06). An 11 year old girl with chronic active uveitis showed a great improvement of the visus. These data suggest that infliximab can significantly and promptly reduce disease activity and improve the quality of life in patients affected by persistently active and refractory JIA. However, more data are needed to evaluate its efficacy and safety as a long term treatment.

11.5 FK-506 (tacrolimus) treatment in severe juvenile idiopathic arthritis (JIA)

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FK-506 safety and efficacy were retrospectively tested in a reduced number of unresponsive patients with JIA

Patients and methods—The treatment of 12 children affected with JIA (4 systemic, 6 polyarticular, and 2 oligo with severe eye disease) was switched from cyclosporin A to FK-506, because of lack of response and/or unacceptable side effects. Median age was 10.45 (6.33 (14.49)), time from diagnosis 4.75 (2.21 (9.7)), and the female to male ratio was 3/1. All patients completed one year of treatment. FK was used at 0.1–0.3 mg/kg to maintain whole blood concentrations