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

B FINCKH, I FOELDVARI, M MEHRWALD, D HUELSEBUS, A KOHLSCHÜTTER
University Children's Hospital, 20246 Hamburg, Germany

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, sulphydryl (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?

PJ HASHKES, DJ LOVELL
Department of Paediatrics, Sieff and Poriah Hospitals, Israel; Rowe Division of Rheumatology, University of Cincinnati, USA

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 IFNγ in the serum of patients with juvenile idiopathic arthritis and systemic lupus erythematosus

D MIHAYLOVA, I BOYKINOV, D NIKOLOVA, N IVANOVA, K LISITCHKI, S STEFANOV, V GENOVA
Department of Rheumatology, University Children's Hospital, Sofia, Bulgaria

We evaluated the levels of interleukin 6 (IL6), tumour necrosis factor (TNF) and interferon γ (IFNγ) 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 IFNγ 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

L STEIN, S CLARK, G FREED
Departments of Pediatrics, University of North Carolina at Chapel Hill, Chapel Hill, NC and The University of Michigan, Ann Arbor, MI, USA

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.

This work was supported by a grant from the Arthritis Foundation.

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

DMC BRINKMAN*, S KAMPHUIS†, P QUARTIER‡, AM PRIEUR‡, R TEN CATE*, M BIERINGS†, NM WULFFRAAT†
**Department of Paediatrics, Leiden University Medical Centre, The Netherlands; †Department of Paediatric Immunology and Haematology, Wilhelmina Childrens Hospital Utrecht, The Netherlands; ‡Department of Immuno-haematology, "Hopital Necker-Enfants Malades", Paris, France*

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.

Unprimed bone marrow was harvested and T cell depleted (in the first 8 using anti-CD2 and anti-CD3 antibodies, in the last 4 using positive CD34 selection). CD34+ ranged from 0.5 to 4 × 10^6 CD34+/kg and residual T cells 0.1 to 25 × 10^4 /kg. The conditioning regimen included antithymocyte globulin (5 mg/kg/day × 4), cyclosporin (50 mg/kg/day × 4) and in all but one patient TBI (4 Gy). The aplastic period lasted 15 to 28 days. Rheumatological follow up at 3, 6, 12, and 24 months showed a marked decrease in arthritis severity as expressed in core set criteria for JIA activity. 6 patients are in drug free, complete remission, 2 in partial remission, and 1 has a mild relapse with oligoarthritis and raised erythrocyte sedimentation rate, treated with low dose prednisone. Five patients had a VZV infection (12 to 18 months after ASCT), 1 child developed an atypical mycobacterial infection during follow up. Transplant related mortality was unexpectedly high: three children died after 12 days, 17 days, and 5 months after ASCT. All had evidence of infection and haemophagocytosis, compatible with a diagnosis of infection associated haemophagocytic syndrome (IAHS), in rheumatology also known as macrophage activation syndrome (MAS). These three children all had active systemic features of JIA just before or during transplant and had received a graft containing <0.3 × 10^4/kg T cells (obtained by positive CD34 selection). The infectious event just before the occurrence of IAHS/MAS, might serve as a trigger for uncontrollable macrophage activation.

Conclusion—ASCT in this severely ill patient group carries a significant risk of developing fatal IAHS/MAS. Less profound T cell depletion, control of systemic disease before transplant and a slow tapering of steroids after ASCT is advised.

7.6 Modern point of view on juvenile rheumatoid arthritis with uveitis

S SALOUGINA, N KUZMINA, A SHAIKOV
Childrens Department, Institute of Rheumatology, Moscow, Russia

During a 10 year period uveitis occurred in 81 of 758 patients with juvenile rheumatoid arthritis (JRA).

Objective—To describe a series of patients with JRA and uveitis.

Methods—The study included 81 patients (68 (84%) girls, 13 (16%) boys) with JRA and uveitis. We studied antinuclear antibodies (ANA) using Hep-2 cells for 33 patients; HLA-DR status was determined for 37 patients.

Results—85% of the children had arthritis before uveitis. The mean age at onset of arthritis was 3.5 years (range 1–10), the mean age at onset of uveitis was 6 years (range 2–15). The mean interval between the onset of arthritis and uveitis was 3.02 years (range 3.5 before arthritis onset to 12.5 years after). In 68% of the patients the diagnosis of uveitis was made within 5 years after onset of arthritis. 93% of patients had mono-oligoarticular onset, but 50% had a polyarticular course. 23.5% had functional disability, class 3–4. 67% of patients had asymptomatic presentation of uveitis. Ocular complications developed in 53%—cataracts 38%, band keratopathy 11%, glaucoma 2.5%. In a study of 33 children with arthritis and uveitis 94% were positive for ANA. 9% had detectable rheumatoid factors. 15 of 37 patients (20%) had HLA-DR8 (p<0.001).

Conclusion—Clinical and laboratory data of our patients suggest that (*a*) JRA with uveitis is a separate nosological form, (*b*) 25% of patients with JRA and uveitis develop serious articular outcome.

7.7 Tumour necrosis factor and its soluble receptors in activation of the autoimmune process in patients with juvenile rheumatoid arthritis

L OMELCHENKO, V CHERNYSHOV, I DUDKA, T LJUDVIK, T POCHINOK, M VODYANIK, V NIKOLAJENKO, E VYKHOVANETS
Institute of Paediatrics, Obstetrics and Gynecology, Academy of Medical Sciences of Ukraine, Ukraine

We have noticed a strange clinical picture for juvenile rheumatoid arthritis (JRA) in patients in comparison with immunological disturbances. Clinical observation, immunological investigation by two colour cytofluorimetry using monoclonal antibodies (IMK kit, Becton Dickinson, USA), and tumour necrosis factor (TNF) and the p55 and p75 soluble tumour necrosis factor receptors (sTNFR) were used in children. Lymphocyte subsets (LS), TNF, and sTNFR were studied in 69 children with JRA, aged from 3 to 14. Disturbances in main LS were found in all children investigated; most of them had decreased levels of lymphocytes in peripheral blood with a background of leucocytosis. This was accompanied by increased relative and absolute levels of CD3+CD4+ cells and activated T lymphocytes, which expressed HLA-DR molecules. It was discovered that increased activity of the pathological process was accompanied by a decreased level of CD3−/CD16+56+ (NK cells) and an increased CD3+CD4+/CD3+CD8+ ratio. Increased activity of the JRA in patients was accompanied also by an increased level of TNF in comparison with sTNFR and a decreased molar ratio of sTNFR/TNF. The tendency for a decreased molar ratio of sTNFR/TNF and an increased CD3+CD4+/CD3+CD8+ ratio may be a reliable index for prognosis of the increased activity of the autoimmune process in children with JRA.

7.8 Nutritional aspects in juvenile chronic arthritis

L RONDINONE*, L GALLITELLI†, F FANTINI*, V GERLONI*
**Chair of Rheumatology of the University of Milan, Centre for Rheumatic Children, Gaetano Pini Institute, Milan, Italy; †Centre of Nutritional Valuation, Niguarda Hospital, Milan, Italy*

Nutritional status and nutrient intake were assessed in 22 children with juvenile chronic arthritis (JCA). Seven patients had systemic, 8 polyarticular, and 7 oligoarticular JCA. Twenty patients received corticosteroids. The mean age at assessment was 8.6 years (range 1.7–16.6). The mean age at disease onset was 3.1 years (range 1–7.9). All anthropometric measurements were below the normal values.

The body mass index (BMI) was lower (but not significantly) in the patients with polyarticular disease (BMI = 13.1) than in those with oligoarticular JCA (BMI = 14.8) and systemic JCA (BMI = 15.13). The BMI was low in patients with disease onset age less than 3.5 years (BMI = 13.7 *v* 15.5) (p=0.08). Seventeen patients had reduced retinol binding protein (RBP) (mean 17 mg/l), 12 patients had reduced serum iron (mean 3.9 μmol/l), 12 reduced intra-erythrocyte GSH (mean 19 U/g haemoglobin), 6 reduced zinc (mean 672 μg/l), 6 reduced plasmatic GSH (mean 214.8 U/l), 6 reduced vitamin A (mean 143 μg/l), 5 reduced selenium (mean 56.6 μg/l), 4 reduced vitamin E (mean 6.82 μg/ml), 4 reduced folic acid (mean 2.7 ng/ml).

In conclusion, JCA causes metabolic and nutritional disorders associated with low BMI, mainly in the polyarticular form and in patients with early disease onset.

7.9 Macrophage activation syndrome in childhood rheumatic diseases: a tertiary hospital experience

S SAWHNEY, P WOO, KJ MURRAY
Department of Rheumatology, Great Ormond Street Hospital, London, UK

Objectives—To review the incidence, precipitating events, clinical features, treatment, and outcome of macrophage activation syndrome (MAS) occurring in a single tertiary level paediatric/adolescent rheumatic unit.

Methods—Retrospective review of cases of MAS from a prospectively collected database of paediatric rheumatic diseases from 1980 to 2000.

Results—9 patients had MAS. 5 with systemic onset juvenile idiopathic (SOJIA), 1 enthesitis related arthritis, and 1 CINCA syndrome. Seven of 99 total patients with SOJIA developed MAS. Mean age of onset of the primary disease was 5.7 years and disease duration before MAS, 4.2 years. All patients had active disease before developing MAS. No drug change was identified as a trigger. Specific infections before MAS were commonly seen in (8/9). 2 of 3 patients with significant renal impairment died. Tables 1 and 2 give details of clinical data and laboratory tests.

Table 1

Clinical data	*No.*
Female:male ratio	8:1
Infection before MAS	8/9
Active disease before MAS	9/9
Typical fever	9/9
Hepatosplenomegaly	8/9
Lymphadenopathy	6/9
Icterus	2/9

Table 2

Lab/outcome	*Result*
Average platelets fall (8)	346→99
Average ESR fall (3)	115→28
Coagulopathy	6/9
Hepatitis	8/9
Haemophagocyt.	4/7
Survival	7/9
Mean recovery	24 days

All patients received high dose steroids (8 IV/one oral)—5 cyclosporin, 2 cyclophosphamide, and 1 antithymocyte globulin.

Conclusions—MAS is a rare, potentially fatal, complication of childhood rheumatic diseases. In our cohort a female predominance was seen, and the syndrome was often preceded by infection. Deranged renal function is associated with a poor prognosis. Bone marrow studies support the diagnosis. Aggressive early treatment is essential.

7.10 An evidence base for uveitis screening in juvenile idiopathic arthritis

C MACHADO, C BENTLEY, V LEE, E GRAHAM, K MURRAY, P WOO, C EDELSTEN
Departments of Rheumatology and Ophthalmology, Great Ormond Street Hospital for Sick Children, London, UK

Objective—To examine independent risk factors for juvenile idiopathic arthritis (JIA) associated uveitis and establish the period of risk for a critical appraisal of the screening programme.
Patients and methods—315 patients with JIA undergoing regular ophthalmological examination, including 165 with chronic anterior uveitis, were enrolled in a single tertiary referral centre. Baseline risk factors for uveitis were established by univariate and multivariate survival analysis. To establish the appropriate length of the screening programme the actual and estimated periods of risk were established for the subgroups of patients with JIA stratified by the risk of uveitis.
Results—The risk of developing uveitis was associated with earlier age at onset of arthritis in months—HR 0.98 (0.98–0.99, p=0.001), type of JIA at onset: polyarticular *v* oligoarticular—HR 0.24 (0.11–0.51, p= 0.001), and antinuclear antibody (ANA) positivity—HR 2.16 (1.30–3.59, p=0.003). The prevalence of uveitis varied from 91% in those with ANA+ oligoarticular JIA presenting before 20 months to zero in those with polyarticular JIA presenting after 70 months. The gap between the onset of arthritis and the diagnosis of uveitis was only associated with the age at arthritis onset. The maximum gap varied from 76 months in those with extended oligoarticular arthritis aged less than 20 months and was less than 15 months in all those developing arthritis after 70 months.
Conclusion—Owing to these results, we can suggest a rational and effective screening programme for uveitis surveillance with shortening of the screening programme in children developing definite JIA between 4 and 13 years.

7.11 Juvenile idiopathic arthritis and related autoimmune diseases

MG ALPIGIANI, M CERBONI, T MUSTICA, A IESTER, R LORINI
University Paediatric Department, G Gaslini Institute, Genoa, Italy

An association between juvenile idiopathic arthritis (JIA) and other autoimmune diseases has been described. Our study aimed at investigating the coexistence of autoimmune diseases (coeliac disease (CD), autoimmune thyroid disease, and type 1 diabetes mellitus) in our patients affected by JIA. We studied 65 patients (41 female and 24 male) with an average age of 13 years and 2 months (range 2–27 years). 41/65 patients had a pauciarticular form of JIA, 13/65 had a polyarticular form, and 11/65 had a systemic form. Only in one female patient was the CD diagnosed about 2 years before the JIA diagnosis. All patients underwent to an autoimmune thyroid screening through anti-thyroglobulin (TGA) and anti-peroxidase (TPOA) antibodies determination with fluorescence enzyme immunoassay (FEIA) and through thyroid ultrasound. The type 1 diabetes screening included OGTT and determination of antibodies to glutamic acid decarboxilase (GADA), insulin (IAA), tyrosine phosphatase (IA-2A) by radiobinding assay and islet cell (ICA) by indirect immunofluorescence (IIF). In all patients without IgA deficiency we looked for CD-specific markers, through the dose of the antigliadin antibodies (IgA-AGA) by FEIA, antiendomysium antibodies (EMA) by IIF and antitransglutaminase antibodies by ELISA assay. 9/65 female patients (14%) showed antithyroid antibodies, in particular: TGA in 3 cases, TPOA in 5 cases, TGA and TPOA in 1 girl. In 3 of these patients, the ultrasound described a dishomogeneous thyroid tissue, that led to Hashimoto's thyroiditis diagnosis. As regards type 1 diabetes, only 2/65 patients showed high levels for GADA. For CD, a male patient had high levels of IgA-AGA and EMA; for this reason the child underwent a duodenal biopsy, which confirmed the CD diagnosis (villous atrophy). Our study showed that the frequency of antithyroid antibodies (9/65, 14%) and CD prevalence (2/65, 3%) are higher in JIA than in the general population; type 1 diabetes markers (islet autoantibodies) are not frequent. Therefore it seems to be advisable to look for thyroid autoimmunity and CD-specific markers in patients with JIA.

7.12 Prevalence of iridocyclitis in juvenile idiopathic arthritis

M CERBONI*, MG ALPIGIANI*, R DE MARCO†, T MUSTICA*, P VITTONE†, A IESTER*
**University Paediatric Department, †Department of Ophthalmology, G Gaslini Institute, Genoa, Italy*

Anterior uveitis associated with juvenile idiopathic arthritis (JIA) is an important cause of visual impairment in children. Our retrospective study aimed at evaluating the prevalence of uveitis in a group of children with JIA. Our study included 171 patients (99 female, 72 male) affected by JIA; the average age at onset of the disease was 6 years and 8 months (range 3 months–16 years and 1 month). Among these patients, 94/171 were affected by a pauciarticular form, 38/171 a polyarticular form, and 39/171 a systemic form. Ophthalmological examination confirmed the diagnosis of anterior uveitis in 17/171 patients with a prevalence of about 10%, which is in accordance with published data. Eye complications were cataract in 7/17 patients, posterior synechiae in 7/17 patients, visual loss with a visual acuity snellen of less than 0.3 in 3/17 patients, band keratopathy in 2/17 patients, and glaucoma in one patient. Analysis of the clinical findings showed that female patients (13/17), patients with the pauciarticular form (15/17), patients with a positive antinuclear antibody test (15/17), and patients with articular disease aged less than 5 years have a higher risk for anterior uveitis. Finally, we considered it imperative, in order to prevent visual loss, that there should be early anterior uveitis diagnosis and an accurate ophthalmological follow up as suggested by Fink[1]—that is, for the pauciarticular form every 3 months for 2 years, then every 6 months for 7 years, and finally once a year; for the polyarticular form every 6 months for 5 years, then once a year; for the systemic form once a year.

1 Fink CW, *et al*. Arthritis Rheum 1980;23:673.

7.13 Systemic juvenile idiopathic arthritis associated with Kikuchi's disease

SKF OLIVEIRA, UBW DESTRI, LCO VASQUEZ, S FERMAN, S ROMANO, FR SZTAJNBOK
Paediatric Rheumatology Unit, Federal University of Rio de Janeiro, Brazil

Necrotising lymphadenitis (Kikuchi's disease) is a benign lymphadenopathy that affects preferentially cervical lymph nodes and can remit spontaneously or by treatment with steroids. The disease can be primary or secondary to other immunoinflammatory diseases and in the medical literature there are reports of its association in 4 adults with Still's disease and in one teenager (14 years old) with systemic JIA. Our aim is to report another case in systemic JIA, in a younger patient, comparing its clinical similarities with the cases reported before. The patient is a girl, daughter of a Japanese couple, whose systemic JIA started when she was 6 years old. On the 15th day of the disease she had a worsening of the clinical status owing to pancytopenia and bleeding disorders compatible with macrophagic activation syndrome (MAS). She responded well to steroids, entering into remission after 3 months. For 5 years she had been asymptomatic until the typical fever of JIA started again and 4 months later a rheumatoid rash, lymphadenopathy, and arthritis were seen. The lymph node biopsy was compatible with Kikuchi's disease and steroids were introduced. Among these 6 patients reported so far, 4 had a Japanese background and 2 of them had symptoms that suggested MAS. Extra-articular manifestations such as fever, lymphadenopathy and rashes were more important than arthritis, which usually followed a pauciarticular course. Five patients had a good response to steroids.

7.14 Cardiac manifestations in systemic juvenile idiopathic arthritis

SKF OLIVEIRA, LG GUEDES, JR CARVALHO, AECL PALHARES, S KNUPP
Paediatric Rheumatology Unit, Federal University of Rio de Janeiro, Brazil

Objective—To describe cardiac disease in the systemic form of juvenile idiopathic arthritis (JIA), including clinical features, laboratory, radiological, electrocardiographic, and echocardiographic findings as well as the course of the disease.
Materials and methods—The medical records of 78 patients with the systemic form of JIA were reviewed. A cross sectional, descriptive study was performed to evaluate the cases that presented clinical evidence of cardiac disease.
Results—Symptomatic cardiac disease occurred in 10 patients (13%). There were 5 recurrent episodes. Isolated pericarditis occurred in nine episodes, myocarditis in five, and perimyocarditis in one. Cardiac disease began before the age of 6 in 73% of the cases and within the first year of JIA in 70%. Fever was the clinical manifestation present in all cases. One patient had perimyocarditis associated with cardiac tamponade. Two children died, one of varicella during myocarditis treatment and the other of pulmonary hypertension. The cardiac disease resolved in the others.
Conclusion—The frequency of symptomatic cardiac disease seen was significant and the manifestations present were in accordance with the literature.

7.15 Neurosensorial loss of hearing in systemic idiopathic juvenile arthritis

SKF OLIVEIRA, AFGS MARQUES, LL CAMPOS, AFG SILVA, M RODRIGUES, FR STANJBOK, ACM MARQUES
Paediatric Rheumatology Unit of Federal University of Rio de Janeiro, Brazil

Hearing loss is a rare complication of JIA. In the medical literature we found neurosensorial hearing loss in only one child with a pauciarticular JIA and in one adult patient with

Still's disease. However, in rheumatoid arthritis, this kind of disorder was seen in 20% of a series of 72 patients, which did not correlate with drugs or a hereditary origin.

Systemic JIA started in the patient when he was 5 years old and followed a severe and persistent polyarticular course. At 7 hearing loss was noted on the left side and was attributed to a salicylate toxicity, but it did not revert after withdrawal of the drug. At 16, still with a persistent articular activity, an acute and intense otalgia developed on the left ear, diagnosed initially as media otitis and treated with antibiotics. Five days later the same symptoms appeared on the right ear. An investigation showed a moderate neurosensorial hearing loss in the right ear and a deep one in the left, bilateral absence of estapedian reflex, and tympanometry with type C curve on the left ear and a type A curve on the right. Treatment with IV pulses of methylprednisolone, followed by oral prednisone brought about remission of the loss/deficit in the right ear but no change in the left. There were no other complications during the past 2 years, and the deficit on the left ear remains unaltered.

Development of hearing loss in a patient with JIA needs a differential diagnosis from among many causes, such as neurosensorial autoimmunity, infections, and drugs ototoxicity. In this case, hearing loss was probably autoimmune in origin. The importance of the case is that it shows that intense treatment shortly after diagnosis can alter the prognosis and may reverse the problem.

7.16 Pulmonary hypertension in systemic juvenile idiopathic arthritis

SKF OLIVEIRA, C DOMINGUES, M RODRIGUES, FA MALTA NETO
Paediatric Rheumatology Unit, UFRJ, Brazil

Pulmonary disease in JIA is usually a benign, extra-articular manifestation, characterised mainly by asymptomatic defects of pulmonary function, pleuritis, or pneumonitis. In the past 35 years, pulmonary hypertension (PH) was reported in only 2 patients (1964 and 1991): one with polyarthritis and high levels of rheumatoid factor and the other with a systemic JIA with a polyarticular disease. Even in adults with Still's disease PH was diagnosed only once, in a 29 year old woman whose PH appeared 2 years after her initial diagnosis (1990). Interestingly, we saw this kind of complication in 2 patients with systemic JIA, leading us to describe the clinical and laboratory manifestations and evolution of this condition.

Our first patient was a white girl whose JIA started when she was 3. PH was diagnosed 8 years later and a fatal course followed 3 months after this diagnosis. The second, a 12 year old boy in his 4th year with a badly controlled disease, complained of dry cough for a month and was eventually admitted into the hospital because in the previous 5 days he had presented exertional dyspnoea, orthopnoea, and tachycardia, receiving a PH diagnosis.

Although primary pulmonary hypertension has been reported in patients with a variety of connective tissue diseases, it is rare in children with JIA, and its pathogenesis has not yet been understood. The absence of parenchymal disease in the lung suggests that the disease is related to vessel involvement. Coincidently, persistent and badly controlled arthritis was a common feature in all patients reported so far. PH in the course of JIA brings a high possibility of a severe and non-responsive disease with a rapidly fatal course.

7.17 Temporomandibular synovitis as a unique presentation of juvenile idiopathic arthritis

G MARTINI*, U BACCILIERO†, A TREGNAGHI*, F ZULIAN*
**Department of Paediatrics, University of Padua; †Department of Maxillofacial Surgery, Hospital of Vicenza, Italy*

Temporomandibular joint (TMJ) disease is quite common in juvenile idiopathic arthritis (JIA). We describe a case of TMJ arthropathy as a unique manifestation of JIA, and its successful treatment.

A 15 year old girl was referred for isolated orofacial pain on mastication, increasing difficulty in eating, and weight loss. Past history was unremarkable. The maximal mouth opening capacity (MOC) was 30 mm. Bilateral crepitations, malocclusion, weakness of the mastication muscles were present. Other joints and eye examination was normal. Erythrocyte sedimentation rate, C reactive protein, and other blood tests (HLA-B27, antinuclear antibodies, dsDNA, rheumatoid factor) were normal. Bone ^{99}Tc scintiscan showed increased tracer uptake on both TMJs. Imaging techniques (orthopantomography, computed tomographic scan, and magnetic resonance imaging (MRI)) showed bilateral flattening of the mandibular condyles, increased synovial fluid, and bilateral synovial hyperplasia. The patient underwent arthroscopic synovectomy followed by triamcinolone hexacetonide (10 mg) injection. A synovial biopsy specimen showed prominent villi with 1–4 layers of synovial lining cells with inflammatory infiltrate. At 6 months' follow up a significant improvement was seen: MOC 42 mm, no crepitation, normal strength of the mastication muscles, weight gain 4.7 kg, bone ^{99}Tc scintiscan normal, MRI: complete resolution of synovial thickening but unchanged bone alterations.

The clinical history and the laboratory findings are suggestive of an isolated localisation of JIA. To our knowledge, this is the first patient with this unusual presentation reported in the literature. The good response observed encourages an early use of synovectomy and intra-articular steroids in TMJ disease to reduce pain, improve jaw function, and prevent further irreversible deformities.

7.18 Intestinal pseudo-obstruction due to amyloidosis in a child with juvenile idiopathic arthritis

L LEPORE, S FACCHINI, C MALORGIO, A VENTURA
Department of Paediatrics, IRCCS Burlo Garofolo, University of Trieste, Italy

A 13 year old boy with chronic juvenile idiopathic arthritis of systemic onset with polyarticular disease was referred to our centre after a 3 month history of nausea, vomiting after meals, diarrhoea, and considerable weight loss (from 30 kg to 22 kg). The patient had been receiving anti-inflammatory and immunosuppressive drugs since the age of 6. Inflammation indices remained always high and the damage to the joints had become progressively worse, confining him to a wheelchair. On admission the child was in poor condition with a distended and hypertympanic abdomen. The laboratory test results showed anaemia (86 g/l), raised inflammation indices (PCR 147 mg/l), erythrocyte sedimentation rate (72 mm/1st h), and hypoprotidaemia (albumin 22 g/l). Bacterial cultures of several stool samples were negative. The patient's general condition worsened over the following days and total parenteral nutrition was started. There were still frequent episodes of vomiting (8–10/day) accompanied by numerous loose bowel motions (3–4/day), and further weight loss (1 kg/day) with severe hypoproteinaemia and hypogammaglobulinaemia requiring daily intravenous replacement. After some days, there was melena accompanied by severe epigastric pain and vomiting of undigested matter; the abdomen was distended and tender without peristalsis. Plain abdominal *x* ray pictures showed air levels; the clinical picture together with the radiological examination pointed to a diagnosis of intestinal pseudo-obstruction. The severity of the enteropathy with accompanying loss of proteins, together with an abnormal intestinal motility in a subject with serious chronic rheumatoid disease, pointed to suspect intestinal amyloidosis, which was confirmed by a histological analysis of a rectal biopsy specimen (Congo positive). Chlorambucil was started at a dose of 0.2 mg/kg/d. Over the following two weeks, the child's clinical condition steadily improved; he put on weight, the vomiting and diarrhoea ceased, total parenteral nutrition was stopped, and he returned to a normal diet.

7.19 Detachment of the anterior cruciate ligament: a potential misdiagnosis using MRI of juvenile idiopathic arthritis (JIA) beginning as gonarthritis

H MICHELS*, L SCHUCHMANN†
**FKH, Department of Paediatric Rheumatology, Neckargemünd, Germany; †Paediatric Rheumatology Clinic, Freiburg/Breisgau, Germany*

Problem—Magnetic resonance imaging (MRI) provides us with excellent images of anatomical structures without exposure of the child to harmful radiation. Nevertheless, the images of this relatively new procedure may be misinterpreted. Here, we report two cases of oligoarticular JIA with an onset as gonarthritis where a "detachment tear of the anterior cruciate ligament" was initially diagnosed using MRI by two independent institutes.

Case 1 (female)—At the age of 8.4 years the patient had fallen on her knees. During the next few days the knee continued to become very painful. Using MRI a detachment of the anterior cruciate ligament was diagnosed. Accordingly, the knee was provided with a resting splint. After five days, a swelling of the left knee and of the right ankle developed. At this time, the erythrocyte sedimentation rate (ESR) was 88/104 and an antinuclear antibody (ANA) titre of 1/1280 was found. One month later when she was seen in a paediatric rheumatology clinic for the first time it was discovered that she had iridocyclitis of both eyes and synechiae in her right eye.

Case 2 (female)—At the age of 9.3 years, after a game of handball, a swelling of the left knee developed. Two weeks later, a MRI led to the diagnosis "detachment of the anterior cruciate ligament". Ten weeks later, when the patient was seen in a rheumatology clinic for the first time, she appeared to have arthritis of both knees and of her right wrist. The ESR was 40/91, ANA titre 1/640 positive, HLA-B27 positive.

Conclusion—MRI is better than all other imaging methods, especially the conventional *x* ray. However, to avoid misdiagnoses a correct technique must be used, and the images should be interpreted carefully by an experienced investigator on the basis of the patient's

history, the clinical symptoms and signs, and laboratory findings. A special pitfall in JIA may be the misinterpretation of gonarthritis as "detachment of the anterior cruciate ligament".

7.20 Prognostic role of antinuclear antibodies in juvenile idiopathic arthritis

M BARDARE, M SCARAZATTI, V MARTINI, A GRASSI, A PETACCIA
Paediatric Rheumatology Centre, I Paediatric Clinic, University of Milan, Italy

Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease of childhood and an important cause of disability; the prognosis is multifactorial and the presence of antinuclear antibodies (ANA) is one of the various aspects. ANA are the best known and important autoantibodies in childhood connective tissue diseases. In this study we evaluated the prognostic correlation of ANA in 316 patients affected with JIA. The prevalence of ANA in children varied according to the different subtypes: pauciarticular JIA 87%, polyarticular JIA 55%, and systemic JIA 61%. Our results (table 3) confirm the literature data.

Table 3

JIA	*ANA positivity (%)*
Systemic (24 patients)	61
Systemic ev poly (38 patients)	68
Polyarticular (56 patients)	55
Pauciarticular (167 patients)	87
Pauciarticular ext (31 patients)	87

Our study showed also another important prognostic correlation between the ANA titre and flare up of arthritis in pauciarticular JIA: in 28 of 31 patients (90%) who had a flare up of disease activity an increase in ANA titre was evident. In the pauciarticular JIA population we found no difference of ANA positivity between the group with iridocyclitis (63 patients) and the group without iridocyclitis (104 patients): 94.5% *v* 83% (p>0.05); on the contrary, ANA levels increased in patients who had an iridocyclitis relapse (74% ANA rise in iridocyclitis relapse). In conclusion, in JIA ANA titre is important for: (*a*) JIA identification subtype; (*b*) arthritis flare up; (*c*) iridocyclitis relapse.

7.21 Importance of the synovial fluid aspiration before injecting intra-articular corticosteroids

I CALVO, L LACRUZ
Paediatric Rheumatology Unit, University Hospital Dr Peset, Valencia, Spain

The joint infiltration with corticoids is a common treatment in patients with juvenile idiopathic arthritis (JIA).
Objective—To evaluate in a prospective study whether the synovial fluid aspiration (arthrocentesis) before the joint infiltration with corticoids influences the duration of the clinical effects.
Patient and methods—The study was carried out in 20 patients (16 female, 4 male) with JIA, aged less than 16 years, who presented inflammation signs in the knee (heat, blush, swelling, and pain). A total of 29 infiltrations were performed with triamcinolone hexacetonide in this joint. We studied two groups: 16 cases with arthrocentesis previous to the infiltration and 23 cases without arthrocentesis. In both groups we noted the number of relapses and the period of time between them.
Results—Table 4 shows the results obtained

Table 4

Group	*Arthrocentesis (n=16)*	*No arthrocentesis (n=23)*
Relapses (%)	18.8	47.8
Mean time between relapses (months)	5.0	3.8

Conclusion—Arthrocentesis before the joint infiltration with corticoids reduces the risk of relapse of the arthritis in children with JIA. It is concluded that joint fluid aspiration should be included in the protocol when injecting intra-articular corticoids.

7.22 Ultrasound of the knee: diagnostic value in juvenile idiopathic arthritis

I CALVO, L LACRUZ
Paediatric Rheumatology, University H Dr Peset, Valencia, Spain

Juvenile idiopathic arthritis (JIA) continues to be the most representative illness in paediatric rheumatology. The knee is the most commonly affected joint and often it is not possible clinically to detect synovial fluid. A knee ultrasound (US) scan has been proposed in the past few years as an effective, less invasive, method for the evaluation of articular damage in JIA.
Objective—To evaluate the efficacy of US in assessing joint knee inflammation in children with JIA.
Material and methods—An US of the knee was performed in 40 children with a diagnosis of JIA, aged between 2 and 16 years. The patients were treated with non-steroidal anti-inflammatory drugs, associated or not with methotrexate or corticoids. A physical examination of the knee was carried out to determine pain, the presence of swelling, and the degree of limitation. US scans were obtained with a Toshiba ultrasound equipped with a 7.5 MHZ linear-array transducer. The knee examination was performed always in the same order: anterior, medial, lateral, and posterior area of the knee with longitudinal and transverse US scan. In the US was evaluated: synovial fluid, synovial proliferation, joint cartilage, Baker's cyst, and tendon-like periarticular structures.
Results—In 30 (75%) patients we found joint effusion (bursa suprapatellar and prepatellar), 24 (60%) with synovial proliferation, 15 (38%) with cartilage thickness, 4 (10%) quadriceps tendonitis, and 3 (7.5%) Baker's cysts. The 6 months' follow up showed persistence of minimum fluid in 7, cartilage thickness in 10, and synovial proliferation in 2 patients.
Conclusion—US has demonstrated accuracy and reliability in the diagnosis and follow up of the knee joint inflammation in children with JIA. However, prospective long term studies are needed to assess the suitability of US for the detection of joint disease during the clinical course of JIA and to correlate follow up findings with response to treatment.

7.23 Genetic component in juvenile idiopathic arthritis

A SAVOLAINEN*, H SÄILÄ*, K KOTANIEMI*, O KAIPIAINEN-SEPPÄNEN†, M LEIRISALO-REPO‡, K AHO¶
**Rheumatism Foundation Hospital; †Department of Medicine, Kuopio University Central Hospital; ‡Department of Medicine, Division of Rheumatology, Helsinki University Hospital; ¶National Public Health Institute, Helsinki*

Objective—To obtain information on the magnitude of the genetic component in juvenile idiopathic arthritis (JIA).
Methods—Multicase JIA families were traced systematically at the Rheumatism Foundation Hospital in Heinola, Finland, over a period of 15 years. About 2000 patients with JIA were seen. (The average number of children in Finnish families is 1.8; 45% of families have only 1 child, 38% have 2 children, 13% have 3 children, and 4% ⩾4 children.)
Results—A total of 41 families with 88 affected siblings were found fulfilling the Durban criteria for JIA. In 60 of the cases (68%) the disease was pauciarticular and, in most instances, it ran a mild course. The mean age at JIA diagnosis was 4.6 years. Over the same period, 8 sets of monozygotic twins were found; two twin pairs were concordant for JIA.
Conclusion—Taking into account the prevalence of JIA (1 per 1000), the figures indicate that the sibling recurrence ratio is fairly high, higher than previously believed.

7.24 Significance of raised serological markers of coeliac disease in children with juvenile rheumatoid arthritis

S M AL-MAYOUF, A ALMEHAIDIB, M ALKAFF, S BAHABRI
Department of Paediatrics, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

Objectives—(1) To determine the frequency of coeliac disease (CD) in a group of children with juvenile rheumatoid arthritis (JRA). (2) To determine the correlation between the presence of the serological markers and the histological diagnosis of CD.
Methods—40 children (24 girls) with JRA, aged between 5 and 15 years, were studied. The diagnosis of JRA was based in all cases on the criteria of the American College of Rheumatology (ACR). Serological markers for CD (gliadin-IgA, gliadin-IgG, reticulin, and endomysim-IgA antibodies) were determined with the standard ELISA (Pharmacia UniCAP-100 system) and indirect immunofluorescence methods. Patients in whom serological markers were detected underwent endoscopic intestinal biopsy. The diagnosis of CD was based on the classic finding of villous atrophy and crypt hypertrophy.
Results—Sixteen patients (40%) had serological markers for CD. Eight with a systemic form, 5 with a polyarticular form, and 3 with a pauciarticular form of JRA. Levels of AGA-IgG were high in 13 patients (81%), 4 patients (25%) had high levels of AGA-IgA and 6 patients (37.5%) had antiendomysim antibodies (AEA). None of the patients had antireticulin antibodies. Fourteen patients underwent intestinal biopsy; only 1 patient with AEA (2.5%); biopsy showed typical finding of CD. The patient with CD showed improvement in both growth and articular symptoms after starting a gluten-free diet.
Conclusion—Our study shows that the screening for silent CD among children with JRA may be useful. Those patients with AEA need further follow up because these antibodies are quite sensitive and specific for CD.

Table 5

	ALL	*ALL JIA*	*ALL (p values)*	*Sensitivity (%)*	*Specificity (%)*
Night time pain	43/62	17/113	0.001	69	91
Early disability	35/62	45/119	0.02	56	62
Blasts	13/59	0/115	0.001	21	100
Low WBC*	9/57	0/103	0.001	16	100
Low platelets†	28/61	5/116	0.001	46	77
Anaemia‡	35/62	38/117	0.002	56	74
Raised LDH	40/54	11/71	0.001	74	84
Raised UA	6/46	2/70	0.028	13	97
Abnormal *x* rays	36/54	7/81	0.001	67	91
Rash	9/62	25/119	0.27	—	—
+ANA	8/36	36/100	0.13	—	—

*Low WBC = WBC $<4 \times 10^9$ cells/l.
†Low platelets = low normal platelets $0.15–250 \times 10^9$/l.
‡Anaemia = haemoglobin <110 g/l.

7.25 Differentiation between acute lymphocytic leukaemia (ALL) and juvenile idiopathic arthritis (JIA)

O JONES, E RABINOVICH, S BOWYER, P DENT, N ILOWITE, C SPENCER, ET AL
La Rabida/University of Chicago, Chicago, IL 60649 and 30 other Pediatric Rheumatology Centers

One of the challenges of paediatric rheumatology is the diagnosis of children with ALL who are referred as possible JIA. We used an internet questionnaire in a retrospective case-control study to obtain predictive factors in 62 children with ALL and 120 children with JIA from 31 international centres. Data analysis was performed using two sided Pearson χ^2 or Fisher's exact tests.
Results—Table 5 shows the results obtained.

Using multivariate analysis and regression modelling, we evaluated further several predictive models for ALL. The best were: (*a*) two of three factors present (1/3 haematological values low, raised lactate dehydrogenase (LDH), night time pain; sensitivity 76%, specificity 86%); (*b*) blasts + 2 of 3 criteria (1/3 haematological values low, raised LDH, night time pain; sensitivity 81%, specificity 86%). These two models, as well as night time pain and raised LDH, and abnormal *x* rays by themselves, may be useful in differentiating a child with leukaemia from a child with JIA and indicating the need for a bone marrow examination.

7.26 ANA detection by recombinant antigens in juvenile idiopathic arthritis and connectivitis

U SANDINI-POHJAVUORI*, V AUBERT†, P ROUX-LOMBARD‡, J-J CHESEAUX*, M HOFER*
*Consultation Multisite Romande de Rhumatologie Pédiatrique, *Departments of Paediatrics University Hospital Lausanne and Geneva; Division of Immunology and Allergy, †Lausanne and ‡Geneva*

In children with juvenile idiopathic arthritis (JIA), antinuclear antibodies (ANA) are frequently positive, in particular in patients with higher risk of uveitis. ANA are classically measured by immunofluorescence (IF), but a new ELISA (ANA-screen) using recombinant antigens has been developed, which may simplify measurement of ANA. Our study aimed at comparing both techniques in children with rheumatic diseases. ANA were measured in 3 groups of children: JIA (A, n=24), lupus and mixed connective tissue disease (B, n=4), healthy controls (C, n=22).

Table 6 shows the results obtained. With the ELISA technique, none of the patients with JIA had positive ANA, though 67% of them had positive IF. In contrast, all connectivitis patients had positive ANA by both IF and ELISA. In healthy controls, ANA were always negative by ELISA, but positive in one patient by IF. Our study suggests that ANA-screen is useful for ANA screening in children in whom connective tissue disease is suspected, but not in patients with JIA. To evaluate if the ELISA technique has a higher specificity than IF a larger control group is necessary.

This work was supported by Pharmacia, CH.

7.27 Health related quality of life (HRQoL) of children with juvenile idiopathic arthritis (JIA)

I FOELDVARI, M REDEGELD, M MEHRWALD, D HÜLSEBUS, M BULLINGER
Paediatric Rheumatology Unit and Department for Medical Psychology, University of Hamburg, Germany

Introduction—HRQoL in children pertains to physical, emotional, mental, social, and behavioural aspects of wellbeing and function as perceived by the patients and their parents. Parents views of HRQoL in patients with JIA are as yet unresearched.
Aim—Differences in HRQoL according to clinical subgroups (for example, diagnosis, morning stiffness) are examined.
Methods—In a longitudinal study the KINDL, a German generic health related quality of life instrument for children, and the Child Health Assessment Questionnaire (CHAQ) were administered to parents of 108 children with JIA. Sociodemographic and clinical data were also collected.
Results—Cross sectional pretreatment data analysis yielded moderate overall scores in both KINDL and CHAQ scales as well as significant parent rated HRQoL impairments for older children, prolonged morning stiffness, and functional problems. Sex, type of disease, and Steinbrocker index did not affect HRQoL status in the parents' view.
Conclusion—Parents rate their children's HRQoL positively. Their rating seems dependent on their children's age, symptoms, and functional status, but not on sex and diagnostic information. Further analysis comparing parent's and children's views of HRQoL are underway.

Table 6

	IF (>1/80)		*ELISA (ANA-screen)*		
	Positive	*Negative*	*Positive*	*Equivocal*	*Negative*
A	16 (67%)	8 (33%)	0	1 (4%)	23 (96%)
B	4 (100%)	0	4 (100%)	0	0
C	1 (5%)	21 (95%)	0	0	22 (100%)

7.28 Serum ferritin (SF) and serum glycosylated ferritin (SGF) evaluation in systemic juvenile rheumatic diseases in childhood

S COMPEYROT*, P QUARTIER*, G LE MOEL†, G CHÉDEVILLE*, AM PRIEUR*
Hôpital Necker, †Hôpital Bichat, Paris, France

SF and SGF levels are considered to be a diagnostic tool in adults with active Still's disease (ASD).[1] In children, syndromes of recurrent fever with systemic symptoms may be difficult to differentiate from systemic juvenile idiopathic arthritis (S-JIA). We thus evaluated the levels of SF and the concentration of SGF in 11 children with S-JIA and in 11 controls. The normal values for SF are 10–200 µg/ml and 50–80% for SGF. We found high levels of SF in 9 children with active S-JIA (mean 2707 µg/ml, range 101–14 500 µg/ml) and low SGF concentrations (mean 10.3%, range 2.8–21%) and normal levels in two children with inactive S-JIA. As expected, normal SF levels were found in 2 patients with systemic lupus erythematosus, 1 with rheumatoid arthritis, and 1 with juvenile dermatomyositis as well as in a child with hepatic tuberculosis who was receiving specific treatment. High levels of SF were found in macrophage activation syndrome (SF 1132 µg/ml, <5% SGF), familial recurrent fever (SF 340 µg/ml, SGF 32%), leishmaniosis (SF 437 µg/ml, SGF 100%), metastatic hemangioendothelioma (SF 252 µg/ml, SGF 13%). In 2 CINCA syndromes, SF levels were in the normal range (77 and 78 µg/ml) but SGF concentrations were <5% in both cases. These preliminary data led us to investigate further the SF and SGF profiles in children with chronic inflammatory diseases associated with high recurrent fever and rash which do not fit with the diagnosis of S-JIA.

1 J Rheumatol 1994;21:890–5.

8 Mechanical and orthopaedic problems

8.1 Aseptic femoral necrosis in a patient with systemic JIA

M TRACHANA*, P PRATSIDOU-GERTSI*, F PAPADOPOULOU†, DE KOLIOUSKAS*, A CHRISTODOULOU‡, J BENIZELOS¶, F KANAKOUDI-TSAKALIDOU*
A' Department of Paediatrics, Aristotle University; Departments of †Radiology, ‡Orthopaedics, ¶Pathology, Ippokration Hospital, Thessaloniki, Greece

A 15 year old girl with a systemic onset of JIA and a persistent systemic course, who had been followed up for 14 months in our clinic, presented with a continuous pain, unresponsive to analgesics, located in the soft tissues of the lower part of her right thigh. Up to that time the patient had received prednisone