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 × 10⁹ cells/l.

†Low platelets = low normal platelets 0.15–250 × 10⁹/l.

‡Anaemia = haemoglobin <110 g/l.

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

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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

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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 recom-

binant 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 connective tissue 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.

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

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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.

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

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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

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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

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%)

(PS) 0.8→0.3 mg/kg/d for 4 months and then on alternate days, methotrexate 15 mg/m² and indometacin 2 mg/kg/d. At the time the pain occurred she was receiving indometacin, PS (20 mg/kg/d), and cyclosporin A (3 mg/kg/d). There was no previous history of trauma in the affected limb. Her imaging work up showed the following: x rays of knees and femur—skip lesions in the right femur, one sclerotic in the middle shaft with heavy periosteal reaction and another one mainly lytic with destruction of the cortical bone and periosteal reaction. Magnetic resonance imaging showed skip lesions localised in the middle third of diaphysis and in the distal metaphysis of the femur. The lesions destroyed the cortical bone, infiltrated the bone marrow, and produced a periosteal reaction without extension to the soft tissues or in the epiphysis. These lesions were indicative of a possible bone solid tumour—that is, osteosarcoma or Ewing's sarcoma. However, a thallium scan was not indicative of malignancy. Her haematological and biochemical findings, including alkaline phosphate and lactate dehydrogenase, were also within normal limits. An open bone biopsy was then performed and ruled out the chance of malignancy; it showed aseptic necrotic lesions in the femur. The case is presented because of the unusual location and imaging appearance of the aseptic necrosis.

8.2 Disablement by untreated systemic juvenile idiopathic arthritis

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Background—We commonly only know the final state of systemic juvenile idiopathic arthritis (sJIA) from pictures in medical textbooks. Nowadays effective drugs exist which can stop the inflammation preventing disability.

We present an 8 years 3 months old girl who had had sJIA since the age of three.

Case report—Personal history: 1994 diagnosis of sJIA. 1995 begin treatment with non-steroidal anti-inflammatory drugs, 1995 additional treatment with steroids. 1996/97/98 tapering of steroids and discontinuation in November 1998. January 1998–99 treatment with growth hormone because of impressive growth retardation. January 1999 exacerbation of sJIA. A trial of treatment with methotrexate was discontinued after two weeks. Since then only homeopathic treatment has been given by the parents. Clinical examination December 1999: bad health, all joints are extremely swollen, very painful on movement and palpation, severe limitation of motion, and functional loss in every joint. The girl can nearly walk; she cannot stand up or sit down or lie down when in sitting position. She is not able to lie flat on her back or to dress or undress, her neck is fixed in a flexion contracture and she can neither look up nor turn her head at all. Growth retardation, retrognathia. x Ray: very severe destruction with loss of joint space, erosions, and signs of aseptic necroses. Possible instability of the atlanto-occipital joint.

Conclusions—Systemic JIA is a severe chronic illness, which untreated can lead to most severe destruction despite its mostly favourable prognosis. In most cases sJIA can nowadays be treated effectively with drugs to prevent disability. Children with sJIA therefore must be treated effectively and in time.

8.3 Pain in arms and legs in childhood

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Objective—To describe the different causes and characteristics of a group of children with referred pain in the arms and legs.

Materials and methods—We studied, from 1995 to 1999, 208 school age children from Tenerife island, with referred pain in arms and legs. The diagnosis was made from a clinical history, physical examination, blood tests, and radiological study in all of them; additional tests were need for some of them (bone gammagraphy, computed tomography, or magnetic resonance).

Results—Of the 208 children, 102 presented organic causes and 106 were diagnosed as growth pains. In the group with organic pains, 38 were diagnosed as arthritis (17 were septic arthritis and 21 juvenile chronic arthritis, 12 oligoarticular, and 9 polyarticular), and 64 as myositis. In the group with arthritis the joints most commonly affected were the knees (left knee in 14 children, and right knee in 6). In the group with myositis 80% of the children had had an enteric or upper respiratory infection previously. In the group labelled as growth pains, additional tests (bone gammagraphy, muscle enzymes) were carried out in 8 children to rule out organic causes. The course was favourable in all the patients with growth pains and myositis. In the arthritis group, treatment was started in all of them and the outcome was favourable in only 29.

Conclusions—Pain of arms and legs, particularly growth pain, in childhood is common, though few reports exist in the current literature. Moreover, they are an important cause of absence from school, causing concern to parents. The results of our study confirm this assumption.

8.4 Cementless total hip arthroplasty in juvenile idiopathic arthritis

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From 1975 to 1997 40 patients with juvenile idiopathic arthritis (JA) had 68 cementless hip replacements. Most prostheses used were Zweymüller prostheses. Autotopic bone grafts were used for important acetabular defects. The average age of the patients at the time of surgery was 17.5 years (range 11–29). JA was systemic in 12 cases, polyarticular in 28 cases. Sixteen hips had had a surgical synovectomy with triamcinolone hexacetonide previously and JA was active in 16 cases. Functional results were evaluated according to the Postel Merle d'Aubigné scale for the hip, and the Steinbrocker classification was used for global function. Mean follow up was 7 years (range 3–15). 6 patients required revision: one for infection, three for acetabular loosening with Ender component, one femoral shaft fracture and a loosening of the Zweymüller acetabular cup owing to a bad primary fixation.

Results for the hips were rated excellent and good for motion and pain. However, functional results observed were quite different because of the location of arthritis, and 8 patients needed another reconstructive procedure to recover good function. A radiographic review showed good bone integration

of the components despite early migration in cases of bad primary fixation. It showed the thickness of the femoral shaft bone around the femoral piece. Use of cementless prostheses appears to be a good solution in this disease, preserving bone stock and showing good radiological and functional results.

9 Rare diseases

9.1 Urinary glycosaminoglycan in the course of FMF

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Familial Mediterranean fever (FMF) is characterised by recurrent fever and serositis. The most important complication of the disease is amyloidosis, which is diagnosed by biopsy. Cheaper and non-invasive methods would be important in the early diagnosis of amyloidosis. We have attempted to study the role of urinary glycosaminoglycan (GAG) in the early diagnosis of amyloidosis. The study group included 123 patients with FMF without attack and 11 patients with FMF secondary amyloidosis. Patients with acute attack were excluded. Eight healthy children and 10 patients with primary nephrotic syndrome served as controls. Microalbumin was also measured in patients with FMF. In patients with amyloidosis, urinary GAG levels were lower than in patients with FMF, those with nephrotic syndrome, and controls. In 49 patients with FMF with a low GAG level, urinary GAG levels increased significantly with incremental increase in the colchicine dose ($p < 0.05$). In some patients with low GAG levels, microalbuminuria was also detected. In these patients, microalbuminuria also decreased along with the increase in urinary GAG, when the colchicine dose was increased. These results suggest that in patients with FMF, monitoring urinary GAG and microalbumin levels may be important in the regulation of the colchicine dose and prevention of amyloidosis. We suggest that effective colchicine dose may be monitored by following urinary GAG levels.

9.2 Epithelial cell-derived neutrophil activator (ENA-78) levels in patients with FMF

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The exact mechanism triggering acute attacks in familial Mediterranean fever (FMF) is unclear. Neutrophil is the effector cell of the inflammatory response at the serosal surface.

Epithelial cell-derived neutrophil activator (ENA-78) a recently discovered chemokine, is one of the most important chemotactic cytokines for neutrophil chemotaxis. We have examined plasma ENA-78 levels in 63 patients with FMF. Thirty one patients had acute attacks and 32 patients had remission. Mean (SD) ENA-78 levels were greater in patients with acute attacks than in attack-free patients (2186.45 (1046.37) v 1646.78 (774.93), $p < 0.05$). The ENA-78 level correlated with erythrocyte sedimentation rate and