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 and have attempted to the style 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) is a recently discovered chemokine, 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 (21.86±45 (1046.37) e 1646.78 (775.93), p<0.05). The ENA-78 level correlated with erythrocyte sedimentation rate and
We aimed at elucidating the role of L-selectin in familial Mediterranean fever (FMF) adhesion molecules in familial Mediterranean fever (FMF) and the velocardiofacial syndrome (VCFS) has recently been described as a new separate group in total of eight cases. We report a further five cases, all of whom have deletions at the 22q11 locus.

Results—All five patients had a history of congenital heart disease (CHD) and velophaRYngeal insufficiency (VPI). 4 have significant learning difficulties. None had a history of recurrent infections but two have a selective IgG deficiency and one has abnormal T cell function with a defective phytohaemagglutinin response. In all patients the arthritis is polycarticular onset or extended pauciarticuLar in nature, progressively requiring treatment with modifying agents and indistinguishable from true JIA. Three of our patients were diagnosed with VCSF in the first years of life as a result of the combination of CHD and VPI. In the other two the syndrome diagnosis was made at 17 and 21 years after rheumatology review. The immunological profiles of the 13 total cases of VCFS with JIA are reviewed: 5 are anti-
clear antibody positive, 3 rheumatoid factor positive, 5 have T cell deficiencies, and 4 (2/5 of ours) have selective IgA deficiency. The increased incidence of both IgA deficiency and JIA in the 22q11 deletion syndrome provides further evidence for possible genetic factors in the pathogenesis of JIA. In addition, the presence of demonstrable T cell defects in 5/13 patients adds further weight to the hypothesis that JIA is a T cell related disease.

Conclusions—The severity of the arthritis in VCFS may reflect underrecognition of milder cases of joint disease in this syndrome and it is important for clinicians to be aware of the association. In addition, the possibility of VCFS should be considered in any patient with JIA in the presence of other features of the syndrome, such as CHD, speech problems, and learning difficulties.

9.4 Effects of colchicine on neutrophil adhesion molecules in familial Mediterranean fever (FMF)

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Introduction—The association between juvenile idiopathic arthritis (JIA) and the velocardiofacial syndrome (VCFS) has recently been described as a new separate group in total of eight cases. We report a further five cases, all of whom have deletions at the 22q11 locus.

Results—All five patients had a history of congenital heart disease (CHD) and velophaRYngeal insufficiency (VPI). 4 have significant learning difficulties. None had a history of recurrent infections but two have a selective IgG deficiency and one has abnormal T cell function with a defective phytohaemagglutinin response. In all patients the arthritis is polycarticular onset or extended pauciarticuLar in nature, progressively requiring treatment with modifying agents and indistinguishable from true JIA. Three of our patients were diagnosed with VCSF in the first years of life as a result of the combination of CHD and VPI. In the other two the syndrome diagnosis was made at 17 and 21 years after rheumatology review. The immunological profiles of the 13 total cases of VCFS with JIA are reviewed: 5 are anti-clear antibody positive, 3 rheumatoid factor positive, 5 have T cell deficiencies, and 4 (2/5 of ours) have selective IgA deficiency. The increased incidence of both IgA deficiency and JIA in the 22q11 deletion syndrome provides further evidence for possible genetic factors in the pathogenesis of JIA. In addition, the presence of demonstrable T cell defects in 5/13 patients adds further weight to the hypothesis that JIA is a T cell related disease.

Conclusions—The severity of the arthritis in VCFS may reflect underrecognition of milder cases of joint disease in this syndrome and it is important for clinicians to be aware of the association. In addition, the possibility of VCFS should be considered in any patient with JIA in the presence of other features of the syndrome, such as CHD, speech problems, and learning difficulties.

9.5 SAPHO syndrome and hemiparesis in a child

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SAPHO syndrome is uncommon in children. It is sometimes associated with inflammatory bowel disease and ophthalmological conditions. We present the case of a child with acute transitory hemiparesis as presenting symptom of SAPHO.

An 8 year old boy was referred for a right facial hemiparesis and painful, warm, and erythematous mass over the right sternoclavicular joint. The past medical history included only primary enuresis. On admission, neurological examination showed decreased strength in the right upper and lower extremities, claudication but normal pain, thermal and light touch sensation. Laboratory tests showed erythrocyte sedimentation rate 47 mm/1 h, while C reactive protein, antinuclear antibody, rheumatoid factor, complement components, and fungal and bacterial, viral and circulatory culture were normal. Standard x ray and computed tomography (CT) of the osteoarticular lesion were compatible with osteopetrosis. Cerebral CT and magnetic resonance imaging (MRI) were negative; MRI of the spinal cord showed thoracic syringomyelic cavity between D5-D6 and D10-D11. An open biopsy of the sternoclavicular mass showed fibroinflammatory tissue with rare lymphocytes and plasma cells. On follow up the neurological problems completely recovered in a few weeks while the sternoclavicular lesion persisted with intermittent symptoms of pain and swelling.

The diagnosis of SAPHO syndrome is supported by the sternoclavicular involvement, the insidious clinical course, and the histological picture. Skin lesions in children can be absent at onset or appear later on during the course of the disease. A neuropathic arthropathy has been implicated on the basis of the level and extent of the syringomyelia, the preservation of pain, thermal sensation and cutaneous trophism, and the histology. However, the coincidence between the onset of SAPHO syndrome and acute transitory hemiparesis is peculiar and not previously reported.

9.6 Puzzling problems for the diagnosis of inflammatory diseases

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The diagnosis of inflammatory disorders remains a challenge for the physicians. Indeed, each separate entity can mimic another one clinically and there are few biological tests to help the diagnosis. Moreover, a number of these diseases may aggregate in the same patient or in the same family.

Case 1—A 6 year old girl with periodic fever syndrome and ulcerative gastroenteritis and vascular purpura (HLA-B51−), one mutation of the gene responsible for FMF (M694V), one M694V mutation in the other gels’ syndrome (HLA-B51+).—An 11 year old Algerian boy with prolonged fever, myalgia, purpura and arthrits (2 MEFV mutations M694I, M694I), final diagnosis: prolonged febrile myalgia syndrome showing FMF.

Case 2—A 3 year old boy with recurrent fever, oral ulcers, oedematous joints, and seronegative arthritis with recurrent fever at the age of 4 years and 7 months. Her fever appeared every week and lasted for 3 days. It was accompanied by cervical lymphadenopathy, abdominal pain, arthralgias, and raised inflammatory proteins and erythrocyte sedimentation rate. Infection and malignancy were excluded. Repeatedly, the IgD level was over 140 mg/l. The patient was treated with prednisone and the fever disappeared. Further studies are needed.

9.7 Hyper-IgD syndrome: case report

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Hyper-IgD syndrome (HIDS) is characterised by attacks of spiking fever, which last for 1–7 days preceded by chills and rigors and accompanied by a number of symptoms—lymphadenopathy, abdominal distress, skin manifestation, arthritis, splenomegaly, etc. In 1999 the gene for mevalonate-kinase was discovered as the gene responsible for HIDS. Our patient is a girl who came to our department with recurrent fever at the age of 4 years and 7 months. Her fever appeared every week and lasted for 3 days. It was accompanied by cervical lymphadenopathy, abdominal pain, arthralgias, and raised inflammatory proteins and erythrocyte sedimentation rate. Infection and malignancy were excluded. Repeatedly, the IgD level was over 140 mg/l.

The patient was treated with prednisone and the fever disappeared. After 4 weeks of steroid treatment the fever appeared again and IgD reached its maximum—198 mg/l.

The patient is now 9 years old, takes 2.5 mg prednisone daily, is without any symptoms, and her IgD concentration remains high.

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Currently, DNA analysis for MVK gene is being performed.

9.8 Chronic joint pain in tricho-rhino-phalangeal syndrome type I

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Tricho-rhino-phalangeal syndrome type I (TRPS) is an autosomal dominantly inherited syndrome characterised by hypotrichosis of the scalp hair, bulbous tip of the nose and skeletal abnormalities including cone-shaped epiphyses of the phalanges and hip.

Our patient is a girl, the first child of three brothers born to healthy unrelated parents. No consanguinity. Gross motor development was slightly delayed, but intellectual development has been normal. She was bald until the age of two. She developed a facial impression of TRPS: the scalp hair was thin and sparse, protruding ears, a long broad philtrum, and a pear-shaped nose. Since reaching school age she has had frequent and often daily complaints of pain in the neck, back, and peripheral joints, especially knees, ankles, fingers, and toes. She has pronounced hypermobility in both small and large joints and laxity of the skin. She has swelling of the proximal interphalangeal joints of both hands with bilateral ulnar deviations in the middle phalanges of the index fingers. Radiological examination of the hands has shown cone-shaped epiphyses of both second indices. Bone age development is retarded. There are no malformations in the hips.

Combined conventional cytogenetic banding analysis and molecular cytogenetic analysis disclosed a pathogenic aberration showing a complex, apparently balanced, translocation t(7;13;8)(p21;q21;q24.1)del8(q24.1).

The breaking point on chromosome 8 (8q24.1) has resulted in an interstitial deletion of at least 3 Mb covering most of the TRPS1 gene region that has recently been cloned.

1 Nature Genet 2000;24:71–4

9.9 X-linked lymphoproliferative syndrome with intracranial and pulmonary aneurysms

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X-linked lymphoproliferative disease (XLP) is an inherited immunodeficiency to Epstein-Barr virus (EBV) infection that has been mapped to chromosome Xq25. The most common presentation is fulminant infectious mononucleosis (IM), but more than 10% of boys have problems, usually infections or lymphoma without EBV infection. Our patient was born in 1985 as the second of three brothers to healthy unrelated parents with negative family history. Both brothers died in early childhood after a short febrile condition resembling FIM. Our patient was well until 12 years when he started to have repeated respiratory symptoms and his x ray showed a stenotic lesion of progressive fibrotic process of both lungs. In November 1999 he was admitted to our hospital and during hyperventilation at functional pulmonary test he suddenly developed massive cerebral bleeding. Subsequent careful imaging showed excessive aneurysms all over his cerebral and pulmonary arterial system. DNA analysis confirmed diagnosis of XLP (mutation of C16ST in the second exon of the gene that causes an amino acid change in position 55—an Arg change to a stop codon). This phenotype of XLP was never seen and we can only speculate about the vasculitic process causing multiple aneurysm changes. The patient is receiving corticosteroid treatment and recovering from the consequences of cerebral bleeding.

9.10 CINCA (chronic, infantile, neurological, cutaneous, and articular) syndrome: report on three new cases

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We describe 3 patients with CINCA syndrome, a rare multisystemic inflammatory disease of unknown cause. In all cases the typical rash was present at or within the first days of life; thereafter the disease progressively affected other organs.

In the first case the rash was associated with papillodema, spiking fever, transient arthritis, and a non-inflammatory lymphadenopathy, identified by biopsy as a reactive lymphadenopathy with mixed hyperplasia.

The second case showed recurrent arthralgia and patellar hypertrophy. At direct immunofluorescence examination, the skin biopsy showed an urticaria-like vasculitis affecting the small and medium vessels of the dermis with IgM deposits in the dermis-epidermis junction.

The third case was characterised by transient arthritis, perceptive deafness, patent oval overgrowth, papillodema, morphological facial modifications, and spiking fever. Skin biopsy showed a leucocytoclastic vasculitis with IgM and complement deposits.

None of our cases had mental retardation (though described as a constant feature).

In all 3 cases laboratory findings showed leucocytosis, increased serum immunoglobulins, and raised C reactive protein and erythrocyte sedimentation rate. In 2 cases the neutrophil activation mark- ers were studied: CD11b and CD18 being greatly raised. These surface antigens have a role in the production of interleukin 8 and in the response to this cytokine. Chronic activation of both circulating and tissue neutrophils, suggesting a primitive defect of this cell line and of osteoclastic cells, which share the same staminal cells, may have an important role in the pathogenesis of CINCA.

9.11 Primary Šiögren’s syndrome (pSS) in children and adolescents: clinical, immunological, and immunogenetic characteristics

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We followed up five girls with average age 14.6 years (range 14–15). All patients had recurrent infections of upper airways, sustained fever, and rash. Systemic symptoms included arthralgia (5/5), Raynaud’s Phe- nomenon (3/5), dental decay (3/5), hair loss (3/5), abdominal pain (3/5). Only two patients had sicca syndrome, detected by Schirmer’s test in 3/5 of the patients. None of the patients had parotid secretion. Immunologically, we found polyclonal hypergammaglobulinaemia (4/5), positivity of rheumatoid factor (3/5), presence of antinuclear antibodies (3/5), anti-Ro/La antibodies positivity (3/5). Immunogenetic HLA typing showed that three of five patients had antigens B8/DR3, clinical and biological changes typical for Sjögren syndrome. Diagnostic criteria for adult pSS are not fully applicable in children and adolescents, because laboratory autoantibody positivity in these patients precedes signs of sicca syndrome. Although anamnestic data, clinical symptoms, and immunological changes may suggest a diagnosis of pSS, sialography may be decisive for the diagnosis in childhood.

9.12 Primary juvenile Šiögren’s syndrome: a rare disease in childhood

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Primary Šiögren’s syndrome is rare in childhood. The disease affects mainly older women and only a few cases of primary Šiögren’s syndrome in children have been described.

We report the case of a five year old girl, whose initial manifestation of disease was Raynaud’s phenomenon. After 6 months, arthralgia and peripheral polyarthrits developed. Two years later recurrent swelling of the parotid glands (mainly right side), dry cough, and vasculitis were additionally seen, and rheumatoid factor and antinuclear antibodies (ANAs) were negative. Raynaud’s phenomenon and antinuclear antibodies were negative. Histological examination of the skin of parotid showed a nonspecific chronic inflammatory cell infiltrate. Laboratory analysis showed a leucocytoclastic vasculitis. We made a clinical diagnosis of primary Šiögren’s syndrome.

We think that it is important to consider primary Šiögren’s syndrome as a possible diagnosis in children with non-specific inflammatory diseases, especially in cases of recurrent fever, rash and arthritis. A careful search for antinuclear antibodies and a biopsy of the parotid gland may help to establish or exclude primary Šiögren’s syndrome as a diagnosis in children.

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antiphospholipid syndrome is usually mild. If the clinical syndrome occurs in a patient with systemic lupus erythematosus, or less commonly in other disorders, such as systemic sclerosis, rheumatoid arthritis, or Behçet’s syndrome, antiphospholipid syndrome is regarded as secondary. In primary antiphospholipid syndrome there is no evidence of other underlying disease.

We report the case of a 14 year old boy, whose initial manifestation of disease was arterial occlusion. Remission of major vessels of the legs with claudication and gangrene of digits. After several months, transient ischaemic attacks, due to thrombosis of intracerebral arteries, developed. The patient conformed to the American Rheumatism Association criteria for the classification of systemic lupus erythematosus with the presence of antiphospholipid antibodies in serum. Good results were achieved with long term treatment.

Upon treatment with an anti-TNF antibody

tumour necrosis factor

and Crohn’s disease, characterised also by an

further and had to be transferred to the

months. The patient was treated with IVIG, phagocytosis. Remission was induced by

our hospital with a first episode of haemor-
sive inflammation; it can occur as a familial

acterised by inappropriate macrophage acti-

CINCA syndrome.

Case report—A 14 year old girl with CINCA presented with large skin ulcers of the legs resistant to conventional local treatment. On biopsy a vasculopathy affecting all layers of the dermis was found, implicating a second-

ar obstruction of the blood vessels, which in

turn may have caused the necrosis. We

suggested that stimulation of angiogenesis

might be valuable in healing of the lesions. The

view of the known ability of PGE1 to stimu-

late angiogenesis in animal experiments, the

patient was treated with continuous PGE1 infusion of 6 µg/kg/24 h for 5 days. This was

was followed by marked improvement in wound

fusion of the lesions and wound repair, which

 persisted for 3 weeks. After this period, the

skin defects deteriorated and a second course of PGE1 infusion was performed, again showing improvement in wound healing. After 1 year of spaced PGE1 courses (4 weeks), permanent closure of the skin defects was seen. Side effects of treatment were pain and erythema on PGE1 infusion.

Conclusion—In this girl with CINCA syn-
drome severe skin necrosis developed, which

might be a consequence of the vasculopathy. The effect of intermittent PGE1 on healing of the skin lesions may include inhibition of platelet aggregation, vasodilatation, and/or angiogenesis.

9.16 Mistakes of, and obstacles to, recognition of neoplastic and haematological disorders mimicking juvenile chronic arthritis

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The present paper is the summary of our observations on neoplastic and haematological disorders that developed with pronounced rheumatological manifestations and were not identified on early stages.

The most rare case of Ashkin’s tumour (from PNET group malignancies) in a 6 year old girl was presented as systemic juvenile rheumatoid arthritis (JRA) (so-called subsep-
sis allergica) followed by vasculitis-like dis-

ease, suspected to be Wegener’s granuloma-
tosis. Ashkin’s tumour was diagnosed after death by histopathological and immunohisto-

chemical analysis, though oncological exami-
nation was done repeatedly during the course of disease with negative results. We also dealt with a case of non-Hodgkin’s lymphoma (Ki-

1), presenting as a vasculitis-like syndrome with asymmetrical polyarthritis in an 11 year old girl. We observed a series of different neoplastic and haematological disorders mimicking juvenile spondyloarthropathies (JSA) owing to the localisation of peripheral

arthritis, the presence of enthesitis, sacroiliitis and axial disease (1 melanoma, 1 mediastinal neuroblastoma, 1 Ewing’s sarcoma, 2 acute leukemias, and 2 patients with heritable spherocytic anemia). One more case of JSA mimicking was found in a teenage boy who had back pain due to exostosis of the lumbar spine (not detected previously by x ray) and talalgia accompanied by ankle arthralgia due to an orthopaedic problem. We also observed 3 cases of secondary knee monarthritis due to atypical localisation of osteoid osteomas, exostosis, and haemangioma.

In all reported cases the mistaken diagno-
sis was caused by (a) pronounced rheuma-

tology manifestations; (b) the confusion of different disorders under a single title; (c) an overestimate of the negative results of the previous diagnostic procedures. Our experience proves that in all doubtful cases differential diagnostic research should be continued.

9.17 Familial Mediterranean fever (FMF) presenting with unusual musculoskeletal manifestations

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FMF is characterised by recurrent episodes of peritonitis, pleuritis, and synovitis. Although the most common musculoskeletal manifestation of the disease is acute recurrent monarthritis other manifestations have been described, including chronic joint disease, spondylarthropathy, myopathy, and the “fe-

brile myalgia syndrome”. The diversity and non-specificity of these clinical features are often an obstacle to the diagnosis of FMF. We describe a group of patients who displayed a variety of non-specific musculoskeletal symp-
toms and in whom genetic screening showed homozygosity for the FMF gene.

Ten patients were Sephardic Jews and 3 were Israeli Arabs. Nine were homozygous for the M694V mutation and the rest were homozygous or compound heterozygous for one of the other 4 mutations (V726A, M680I, M694I, E148Q). Six patients had the “febrile myalgia syndrome”, 2 had recurrent episodes of calf pain and pretilial swelling, 2 had non-
specific myopathy, 1 had recurrent episodes of thigh swelling, 1 had chronic knee arthritis without any other features of FMF, and 1 had spondylarthropathy.

Conclusions—Our observations indicate that genetic screening for FMF should be included in an investigation of recurrent or unexplained episodes of musculoskeletal symptoms among children of Mediterranean extraction.

9.18 Treatment of hyper-IgD syndrome: a question unanswered

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A long term follow up of a child affected with hyper-IgD syndrome (3 years) is reported, with particular emphasis on the treatment of this disease. A male child, born on March 1996 from unrelated, healthy parents, devel-

oped recurrent fever spikes associated with chills, severe malaise, short term diarrhoea from December 1996. Between disease flare
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 (19.45 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.

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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Department of Rheumatology, Gastroenterology and Pathology, G Gaslini Institute Genova Italy 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 mononeric 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 mononeric 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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Our—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 a contemporaneous and discordant result was rescored 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 images. 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.

Although LS is a chronic and progressive disease, in a minority of patients it may have a fluctuating course. Advanced imaging allows an objective assessment of disease activity in LS.

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 extended 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 nif edipine capillaroscopy were normal, except for a high antinuclear cell 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, cranial asymmetry at x ray examination, microvascular ischaemic encephalopathy at MRI, negative cerebral arteriography).

Recently, facial hemiatrophy with a contralateral hemisynthesis 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.