with periodic fever and a high serum IgD, mevalonate kinase was genetically and enzymatically normal, ruling out Dutch-type periodic fever.

Conclusion—Children with unexplained recurrent fevers should be tested for mevalonate kinase deficiency irrespective of serum IgD. Molecular diagnosis can positively identify patients with Dutch-type periodic fever and should serve as a starting point for future trials.

1.24 RAG1+RAG2 expression in peripheral B and T cells of patients with SLE

HJ GIRSCHICK*†, T NANKI†, A GRAMMER†, PE LIPSKY†

*Children's Hospital, University of Wuerzburg, Germany; †Simmons Arthritis Research Center, University of Texas, Dallas, USA

To analyse the potential for receptor replacement the expression of RAG1 and RAG2 mRNA was assessed in the peripheral blood B cell populations and individual B cells obtained from patients with active systemic lupus erythematosus (SLE). Analysis of bulk mRNA by reverse transcriptase polymerase chain reaction (RT-PCR) showed RAG1 and RAG2 expression in peripheral B cells of patients with SLE, but not in normal subjects. Individual SLE B cells, assessed by single cell RT-PCR, showed 3 times more RAG1 and RAG2⁺ cells than normal subjects. 35–65% of double RAG⁺ SLE B cells expressed IgD mRNA, 50% VpreB mRNA, but not IgG mRNA. These results indicate that the enzymes necessary for receptor replacement are present in peripheral IgD and VpreB⁺ SLE B cells. These cells might represent premature bone marrow immigrants. 3% of peripheral SLE memory T cells were RAG1 and RAG2⁺, whereas none was positive in normal subjects. The frequency tripled after αCD3 stimulation in vitro. SLE memory T cells, like B cells, may have the ability to edit their T cell receptor in the periphery.

Supported by the DFG and the NIH, Grant AI 31229.

2 Classification of juvenile idiopathic arthritis

2.1 Evaluation of classification criteria for juvenile idiopathic arthritis (JIA)

M LANGKAMMERER, R HÄFNER, R MORHART

Paediatric Rheumatology Hospital, Garmisch-Partenkirchen

The new classification of JIA means a valuable contribution for international exchange. The definition of distinct categories enables studies of homogeneous groups of patients in the future. Our task at present is to validate the classification criteria.

We therefore categorised a total of 172 consecutive patients with JIA who were treated in our hospital in October 1999.

Results—145 patients fulfilled criteria for one of the defined categories: 30 systemic JIA, 47 oligoarthritis (19 of them with extended oligoarticular course), 44 rheumatoid factor (RF) negative and 5 RF positive polyarthritis, 11 enthesitis related arthritis, and 8 psoriasis arthritis. 27 were classified as “other arthritides”. 11 did not fulfil criteria for any one category, 16 fulfilled criteria for two categories. Double classification occurred mainly in patients with psoriasis or enthesitis.

Discussion—(1) 6 of the 11 patients who did not fit into one category could be classified as oligoarthritis, but a family history of psoriasis

excluded them and criteria for psoriasis arthritis were not fulfilled.

(2) 5 of the 16 patients who met criteria for two categories could be classified in one (psoriasis arthritis) if psoriasis is an exclusion for oligoarthritis, RF negative polyarthritis, and enthesitis related arthritis. The other 11 patients had enthesitis and arthritis but also fulfilled criteria for either oligoarthritis or RF negative polyarthritis.

(3) 25 children categorised as RF negative polyarthritis had asymmetric arthritis of 5 to 8 joints at onset (18 of them with antinuclear antibodies, 7 with chronic uveitis), resembling oligoarthritis. We therefore suggest an extra category “extended oligoarthritis”.

Conclusion—About 85% of our patients with JIA could readily be classified into one of the defined categories. With minor changes in the definition this percentage would increase to over 90%. Introduction of a further category for extended oligoarthritis with asymmetric arthritis of 5 to 8 (9) joints at onset should be considered.

2.2 Classification of juvenile idiopathic arthritis: suggestions to improve the efficacy of the Durban criteria

FLAVIO FANTINI, VALERIA GERLONI, MAURIZIO GATTINARA, CRISTINA ARNOLDI

Chair of Rheumatology of the University of Milan, Centre for Rheumatic Children, Gaetano Pini Institute, Milan, Italy

The Durban Classification Criteria for juvenile idiopathic arthritis (JIA) were tested on a cohort of 683 patients (220 male, 463 female) with juvenile onset primary chronic arthritides recorded in the database of our centre. 214 cases fell into the category of “other arthritis”. To reduce the number of cases in this category, two ways were suggested to (a) reduce the exclusions and (b) to consider a hierarchy of the categories.

According to the data of our patients, the exclusions which most commonly hinder an obvious classification are (a) family history of psoriasis in at least one first or second degree relative (for oligoarthritis); (b) psoriasis confirmed by a dermatologist in at least one first or second degree relative (for enthesitis related arthritis); (c) oligoarticular onset for rheumatoid factor (RF) positive polyarthritis.

The hierarchy principle states that if a patient fits into more than one category, he or she should be attributed to the category with the highest rank. In attributing ranks, two general principles might be taken into account: (a) the more severe form should be preferred to the less severe one; (b) the more differentiated form (that is better biologically and/or clinically characterised) should be preferred to the less differentiated one. According to our data the most useful ranking might be: (1) systemic arthritis; (2) RF positive polyarthritis; (3) psoriatic arthritis; (4) enthesitis related arthritis; (5) oligoarthritis; (6) RF negative polyarthritis.

Table 1 Patients classified according to the Durban criteria

	Oligo	Poly RF (−)	Poly RF (+)	Sys	ErA	Pso	Other
paJCA (n=59)	47*				1		11
poJCA (n=25)		25					
po-RF+ (n=2)			2				
sJCA (n=18)				18			
JSp (n=17)					13	1	3
PsA (n=4)		2				2	

*35 with persistent oligoarthritis and 12 with extended oligoarthritis

If the hierarchy principle were accepted, slightly modified Durban criteria would classify almost all patients with JIA and each category would maintain a fruitful and valuable homogeneity.

2.3 Evaluation of Durban Classification Criteria for juvenile idiopathic arthritis (JIA) in Spanish children

R MERINO*, J DE INOCENCIO†, J GARCÍA-CONSUEGRA*

*Hospital Universitario “La Paz” y †CS Estrecho de Corea, Madrid, Spain

Objective—To test the proposed Durban Classification Criteria for juvenile idiopathic arthritis (JIA) in a cohort of Spanish children. **Methods**—125 patients seen between January 1999 and March 2000 were classified according to the following criteria: EULAR for pauciarticular (paJCA), polyarticular (poJCA), poJCA RF positive (po-RF+), and systemic (sJCA) arthritis; Vancouver for psoriatic arthritis (PsA); and ESSG for juvenile spondyloarthropathy (JSp). Patients were reclassified according to Durban criteria, and their clinical/serological characteristics were reviewed.

Results—Table 1 shows the results obtained. **Discussion**—Application of Durban criteria results in a significant number of unclassified patients. According to these criteria, 14/125 of our patients (11%) fell into the “other arthritis” category, 13/14 because of a positive family history of psoriasis.

Interestingly, a 2nd degree family history of psoriasis does not fit the “psoriatic arthritis” criteria, but allows classification of patients into the “poly RF negative” category.

No differences were found in any variable studied between patients classified as “oligo” (n=47) and those with oligoarticular onset included in “other arthritis” group (n=11), except for a family history of psoriasis.

2.4 JCA in the Nordic countries; a population based multicentre study of construct validity of ILAR, in comparison with EULAR classification criteria

LILLEMOR BERNTSON, FOR THE NORDIC STUDY GROUP OF PAEDI-ATRIC RHEUMATOLOGY

Department of Paediatrics, University of Göteborg and Falun's Hospital, Sweden

Background—Childhood chronic arthritis is a heterogeneous group of disorders. New classification criteria (ILAR), for “juvenile idiopathic arthritis” (JIA), have been proposed. They are more descriptive than those formerly used (ACR-JRA, EULAR-JCA) but need validation under “appropriate epidemiological and statistical direction against classifications already in use”.

Objective—To compare construct validity of ILAR and EULAR classification criteria in a cohort of prospectively collected incidence cases of “juvenile chronic arthritis” within 15 defined areas of Sweden, Finland, Norway,

Denmark, and Iceland. The cases were collected during one and a half years, beginning 1 July 1997.

Method—Clinical variables (including those constituting the EULAR and ILAR criteria) and laboratory findings were registered longitudinally for each patient by paediatricians experienced in paediatric rheumatology. A database was constructed specifically for this purpose. In this study clinical findings and laboratory data from the first 6 months (for certain variables also for the first 12 months) were considered.

Results—322 patients qualified for classification according to the ILAR criteria. Of those 322 patients, 291 had a disease duration of >3 months and were classified according to the EULAR criteria. Thus, 31/322 (10%) children were diagnosed only according to the ILAR criteria. 15% of the patients did not fulfil ILAR criteria for any of the categories and 6% fulfilled criteria for more than one category. As an example of the effect of different classification systems in our study, 160/322 were classified as oligoarticular by ILAR criteria and 184/291 by EULAR criteria.

Conclusion—The qualifications for the ILAR criteria are different from those of the EULAR criteria, which makes simple comparison neither possible nor scientifically relevant. One possibility is to study the construct validity of the respective criteria to give us a better idea of the classification and its consequences.

2.5 Juvenile spondyloarthropathies: checking criteria for diagnosis and classification at the start and after 6 months of disease

J DEHOORNE, B VANNEUVILLE, R JOOS, J VANDE WALLE, EMVEYS
Centre for Paediatric Rheumatology, University Hospital, Gent, Belgium

Classification of juvenile spondyloarthropathies remains a "critical" point in paediatric rheumatology.

Objective—To validate classification methods in a university hospital treated population by a retrospective cross sectional study.

Methods—Four sets of criteria especially designed for juvenile patients (Garmisch-Partenkirchen-JSA, SEA syndrome, enthesitis related arthritis, atypical spondylarthritis) and two sets of criteria for patients without age specification (ESSG, Amor) were checked in a group of 20 patients at the start of disease and after six months.

Statistics—The sensitivity and specificity were calculated as well as the reproducibility after six months.

Results—The preliminary results show equal sensitivities for all sets except for the SEA syndrome criteria (which was the lowest). Although in some patients a switch to other types of juvenile chronic arthritis occurred, the initial fulfilment of the criteria was highly predictive for the fulfilment after six months.

3 Connective tissue disorders

3.1 Analysis of Raynaud's phenomenon in an infant and teenage population

ML GÁMIR, MJ RUBIRA, CM VÁZQUEZ, M REVENGA, R HORTAL

Rheumatology Service, Hospital Ramón y Cajal, Madrid, Spain

Objectives—To analyse the onset and evolution patterns as well as the prognostic value of Raynaud's phenomenon (RP) in a cohort of

children diagnosed and followed up in our paediatric rheumatology service.

Material and methods—In a prospective study several clinical, analytical, and outcome variables were collected from a group of patients under the age of 18 years who were admitted to our department because of a history of positive RP.

Results—54 patients (47 female, 7 male) with a mean age of onset of the disease of 13.5 years (range 2–18) were studied. In 31 patients (57%) RP occurred in isolation and in 23 patients (43%) it coexisted with arthritis, subcutaneous nodules and/or hand oedema.

After an average evolution period of 7 years (range 1–14) 10 patients (19%) developed a defined connective tissue disease (CTD) (4 mixed connective tissue diseases, 3 scleroderma, 2 systemic lupus erythematosus, 1 juvenile chronic arthritis), 31 patients (57%) presented some feature of undifferentiated CTD (arthritis, hand oedema, nodules, finger ischaemia, livedo, mucose ulcers, iridocyclitis, positive antinuclear antibodies (ANA), and pathological capillaroscopy pattern). 13 patients (24%) remain currently as primary RP.

Conclusions—(1) Over 50% (75% in our case) of patients with paediatric onset RP, eventually develop a CTD or present features consistent with this kind of disorder.

(2) The greater proportion found in our study might be explained by the selection of patients examined.

(3) Arthritis, hand oedema, co-occurrence of ANA and the presence of findings in nail capillaroscopy, have a good prognostic value.

3.2 Severe cardiac disease among children with diffuse cutaneous systemic sclerosis (SSc) and polymyositis: report of 4 cases

P QUARTIER, D BONNET, JC FOURNET, C BODEMER, PH ACAR, M OUACHÉE, AM PRIEUR
Immuno-hématologie, Cardiologie and Dermatologie pédiatrique, Hôpital Necker-Enfants malades, Paris, France

Patients and methods—We retrospectively reviewed the charts of 4 girls who met the ACR criteria for diffuse cutaneous SSc and had polymyositis, as defined by the presence of proximal muscle weakness and raised serum creatine phosphokinase level. Disease duration and age on admission ranged from 4 to 12 months and from 7 to 13 years respectively. Dysphagia and oesophageal dysmotility were present in 3 patients, arthritis in 2, lung restrictive syndrome in 2, and echocardiographic anomalies in 2, including marked non-obstructive cardiomyopathy (NOCM) in 1 child. One patient secondarily developed severe NOCM and bowel disease.

Results—Treatment with cyclosporin, methotrexate, and corticosteroids (including pulsed intravenous methylprednisolone in 3 cases) improved the skin thickness score, muscle weakness, arthritis, and lung restrictive syndrome. Oesophageal dysmotility and bowel disease did not improve. The 2 patients who had severe NOCM before the triple combination treatment died after 19 and 31 months. Severe NOCM developed in a third patient 6 months after treatment onset. Cardiac transplantation was performed and the child is alive and well 3 months after. One child treated with cyclosporin only is alive after a follow up of 48 months.

Conclusions—As previously described in adults, children with SSc and myositis may be particularly prone to develop severe NOCM.

Earlier and more intensive treatments are probably required. In the case of yet constituted severe NOCM, heart transplantation should be considered.

3.3 Diagnosis of Sjögren's syndrome in childhood

AG CLEARY*, JA SILLS*, E BAILDAM†, JE DAVIDSON*

*Royal Liverpool Children's Hospital, UK;

†Royal Manchester Children's Hospital, UK

Sjögren's syndrome is rare in childhood. We have recently seen 5 children with definite and two with possible Sjögren's syndrome whose investigations highlighted some of the difficulties in diagnosis in the paediatric age group.

Of the children with definite Sjögren's syndrome one had mixed connective tissue disease (MCTD), one juvenile dermatomyositis, and one systemic lupus erythematosus. Of those with possible Sjögren's one has MCTD. The others have no definite underlying diagnosis.

We used the European classification of Sjögren's syndrome as modified by Vitali *et al* 1993¹ to assess these patients and rapidly became aware of difficulties in using these criteria in children.

Four had symptoms of dry eyes, and six had dry eyes on ophthalmological testing. Three have had symptomatic dry mouths and two have had low measured basal salivary flow rates. There are, however, no validated age reference standards for tear and salivary flow rates in childhood.

Sialography, scintigraphy, and histology are used in the classification. Lip biopsy is unacceptable to most children. One patient had a lip biopsy, which proved unhelpful. We have found ultrasound to be well tolerated and to give information comparable with that obtained by magnetic resonance scanning or sialography. Parotid scintigraphy was performed in two children and thought to be abnormal, but interpretation was hampered by the lack of reference standards in childhood.

Investigation of children with possible Sjögren's syndrome in our units has highlighted problems with applying the current diagnostic criteria in childhood. There is a need for age appropriate criteria to be validated for use in childhood.

1 Vitali C, Bombardieri S, Moutsopoulos HM, Balestrieri G, Bencivelli W, Bernstein RM, *et al*. Preliminary criteria for the classification of Sjögren's syndrome. Results of a prospective concerted action supported by the European Community. *Arthritis Rheum* 1993;36:340–7.

3.4 Life experiences of adolescents living with systemic lupus erythematosus

CHM SILVA, W MACHADO, MHB KISS
Department of Paediatrics, School of Medicine, Federal University of Uberlândia and University of São Paulo, Brazil

Systemic lupus erythematosus (SLE) is a severe chronic disease commonly associated with severe personal, social, and familiar repercussions mainly in adolescents. To get to know the way in which these patients perceive and compare themselves according to the changes and the treatment the disease imposes on their routines, monthly meetings with adolescents with a diagnosis of SLE without central nervous system disease have taken place. Identification of the interests of the group of adolescents was possible through discussion and psychodrama techniques conducted by a doctor and by a social assistant with a degree in psychodrama. The