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PAEDIATRIC RHEUMATOLOGY
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LETTER TO THE EDITOR

Palmar Plantar Hyperkeratosis—A Previously Undescribed Skin Manifestation of Juvenile Dermatomyositis

SIR—We report a presentation of juvenile dermatomyositis (JDM) hitherto undescribed—palmar plantar hyperkeratosis—similar in appearance to keratoderma blenorrhagica. The patient later developed other classic skin manifestations of JDM, i.e. heliotrope rash, photosensitive malar rash and Gottron's papules, as well as myopathy, all of which responded to steroid treatment.

An 11-yr-old girl presented with a 2 yr history of rash, 9 months of weakness and arthralgia, 2 months of nasal speech, and recent onset of dysphagia and stress incontinence.

She did have a past history of skin problems which may not be related to the current diagnosis. At the age of 5 yr, she developed a patch of alopecia over the scalp which spontaneously resolved over a couple of months. She also had mild acne over her chest and face that had improved with time. At 6 yr, she developed Sutton's halo naevi on the skin over her back and shoulders. There was no family history of skin problems nor connective tissue disease except for eczema in a paternal cousin.

At the age of 9 yr, after an active summer holiday, the skin over her knees and elbows, and subsequently hands, feet and ankles, became dry and scaly. Reddish, itchy, thickened patches developed over the next few months. In particular, her palms and soles became grossly thickened, fissured and painful, and she would be found awake at night clawing at her severely scaly, itchy and bleeding feet. Her soles became hyperpigmented and were almost black in colour. She was seen by dermatologists who thought she might have eczema or a form of genetic ichthyosis/keratoderma. She was at this time given 40% urea ointment to apply which resulted in significant improvement in the pigmented hyperkeratosis of her feet, but not total resolution. A skin biopsy was not performed. However, the thickened, erythematous patches over her ankles and hands, especially the knuckles, continued to progress over the next 2 yr. In addition, she became increasingly weak over the following months. She was unable to keep up with her friends walking and had to give up dance because of weakness and arthralgia. Her handwriting deteriorated and she was unable to open door handles. Over the preceding 2–3 months before we saw her, her speech became nasal, and then she developed dysphagia, fluid regurgitation through her nose, and a 1 week history of stress incontinence.

On examination, she had classical features of JDM with a photosensitive malar rash, puffy eyes with heliotrope colouration, as well as Gottron's papules over her knuckles, elbows, knees and lateral malleoli. She also had global muscle weakness and wasting, as well as nasal speech and dysphagia. Unusual features noted were hyperkeratotic scaly itchy palms and soles, and dystrophic nails. She had no features of Reiter's syndrome.

Muscle enzymes were raised: creatine kinase 4498 U/l (normally up to 120), aspartate transaminase 280 U/l (normally up to 35), alanine aminotransferase 270 U/l (normally up to 30), lactate dehydrogenase 3564 IU/l (normally up to 770). MRI was consistent with symmetrical proximal inflammation (showing prominent increased signal in the rectus femoris, vastus medialis, sartorius, gracilis and semimembranosus in the proximal thighs) and muscle biopsy showed myositis (infiltrate of lymphocytes and macrophages with fibre necrosis and regeneration). Antinuclear antibody was positive at 1:2560 with a nucleolar staining pattern, but negative for extractable nuclear antigen (including anti-Jo-1 and Sm). The acute-phase reactants were not raised. ESR was 6 mm/h and CRP 1.1 mg/dl.

She was started on oral steroid treatment with rapid improvement in muscle power, fall in muscle enzymes and resolution of dysphagia, stress incontinence and skin rash, including the palmar plantar hyperkeratosis. The interval between Fig. 1A and B, and between 1C and D, is 4 weeks.

This child with classical features of JDM had an insidious onset with hyperkeratosis of her palms and soles as the only presenting symptom for over a year at least. JDM is known to have a myriad of skin lesions, including localized and generalized follicular hyperkeratosis [1] (which is thought to perhaps be due to erector pili myositis and osteal hyperkeratosis) and generalized spirulosis of pilara pityriases rubra [2] (which produces a linear verrucous spirulosis on the back of the hand). Other features of JDM are usually present concurrently. In adults with DM, paraneoplastic cutaneous syndromes include acrokeratosis, acquired ichthyosis and desquamative circinate erythema [3], but this has not been described in children and our patient did not exhibit any clinical or laboratory evidence of neoplastic disease. Long-term hydroxyurea [4] can also result in dryness, pigmentation, plantar keratoderma and band-like erythema on the dorsum of fingers and toes as in JDM. However, our patient had no such exposure prior to onset of her rash, although she did apply 40% urea ointment subsequently with some improvement to her rash. An adult with

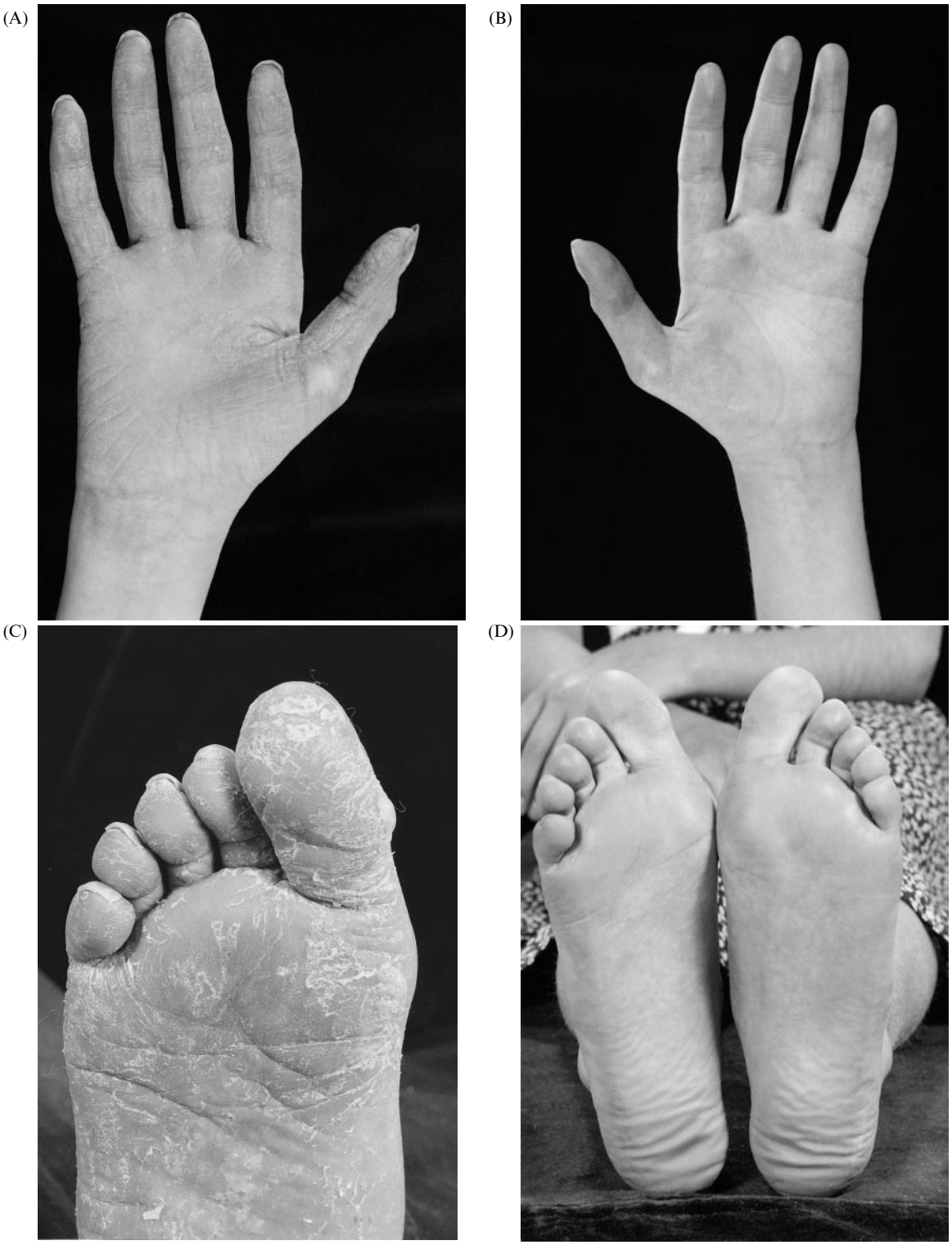

FIG. 1.—(A) Palms before treatment. (B) Palms after treatment. (C) Feet before treatment. (D) Feet after treatment.

'mechanic's hands' and anti-Jo-1-positive polymyositis [5] has also been described. Our patient did not have Jo-1 antibodies.

In summary, hyperkeratosis of the palms and soles in a child can be an early manifestation of JDM.

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INTERNATIONAL LETTER

SWEDISH RHEUMATOLOGY IN GOOD HEALTH AT 50

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THE Swedish model for social security and health care was dependent on a growing economy and full employment. With some 8.5% of open unemployment and faced with growing national debt problems, severe cuts have been implemented in the system and more are to come. How has this affected rheumatologic care? The number of hospital based units remains intact but a general reduction in beds has taken place. Our department in Lund as an example used to have four wards with a total of 62 beds, 12 of which are for day care. This has been reduced to 48, 11 of which are for day care. A complete integration of out- and in-patient services has allowed some staff reductions. However, this has also reduced the total capacity and leads to long waiting lists for out-patient revisits.

As the direct costs paid by patients have increased, health care workers are faced with the novel experience of patients' difficulties to pay medication or afford hospitalization. It should, however, be mentioned that there is an annual maximum of fees for medication of 1300 SEK and services including hospitalization of 900 SEK. One SEK corresponds to approximately 75 pence. However, the most serious threat to the patients is the overall reduction of capacity. In this context new ethical issues have to be faced, and in a typical Swedish way a commission was formed to deal with priorities in health care [1]. This was headed by an emeritus professor of oncology, Jerzy Einhorn, and its strength lies in the fact that all political parties were represented and reached a broad consensus. The highest priority was given to 'care of life-threatening acute diseases and diseases which, if left untreated will lead to permanent disability or premature death', followed by 'care of severe chronic diseases, palliative care and care of people with reduced autonomy' [1].

The universities have not escaped cutbacks but so far this has not affected rheumatology in any major way. The department at Karolinska in Stockholm has, under its new professor Lars Klareskog, undergone a very dynamic rejuvenation and is starting ambitious clinical and experimental programmes. Johan Rönnelid defended his thesis on reactivity to type II collagen and C1q in various clinical situations May of 1997. The resources in Stockholm have now been concentrated to the two teaching hospitals, with Bo Ringertz and Ingiäld Hafström in charge at Karolinska and Huddinge, respectively. The Huddinge group is continuing their research on neutrophil function in inflammation.

In Gothenburg Andrej Tarkowski was appointed as professor in 1996. He has developed a strong academic programme dealing with a new model of septic arthritis, which started with a chance observation by a laboratory technician and which has generated a number of PhD thesis projects, the first was completed by Thomas Bremell, now chief physician of the department. Hans Carlsten is persuing hormonal influences on experimental models of arthritis. Basic T-lymphocyte work is connected with therapeutic investigations, for instance the use of cholera toxin B in induction of oral tolerance. Tarkowski's group has also recently produced good evidence for lymphocyte traffic from the gut to the synovium.

Giant cell arteritis has been studied for a number of years in Gothenburg starting with the thesis of B. Å. Bengtsson and E. B. Malmwall in 1981. The most recent thesis was defended in May 1997 by Christopher Schaufelberger, and included T-lymphocyte cloning and early work on use of T-cell receptors by lymphocytes in peripheral blood and in the lesion. Epidemiologic data showed increased mortality among biopsy negative patients with polymyalgia rheumatica. This disease was also studied in Umeå by Agneta Uddhammar.

In Uppsala, Professor Roger Hällgren has been interested in the study of local events in the gut wall by using a catheter technique allowing local sampling in closed segments. In collaboration with Professor Brandzaeg in Oslo surface immunoglobulins can now be studied in a more precise way. Hällgren has also recently presented data indicating the rationality of administering prednisolone at 2 a.m. rather than in the early morning.

In Lund Tore Saxne's group is now also involved in studies of early events in osteoarthritis and Ingemar Petersson will soon complete his thesis dealing with cartilage and bone derived serum markers in this disease. Of particular interest are results regarding COMP and its relation to clinical, radiological, and other imaging data including scintigraphy. One message will be that bone and cartilage changes both seem to commence early and may be able to distinguish between stationary and progressive phases of the disease. Gunnar Sturfelt's group is involved in SLE studies, making use of a prospective epidemiological cohort and incidence study in a defined population. They find constant annual incidence figures around 5/100 000, and increased mortality in vascular diseases notably early myocardial infarcts in young women. Eva Fex and Kerstin Eberhardt are studying their cohort of rheumatoid arthritis patients. One-third of these patients, despite optimal conventional care,

FIG. 1.—Members of the Department of Rheumatology, in Lund, spring 1997. Photo: F. A. Wollheim.

lose working capacity during the first year of observation. However, on a positive note the remainder continue work over the following 5 years. Radiologic progression is not taking place in erosive patients who on clinical grounds were selected not to receive DMARD therapy. This may indicate that as many as 25% of the initially erosive patients do fare well even without the now so popular early aggressive therapy. ESR/CRP are good predictors.

In Malmö Associate Professor Lennart Jacobsson is interested in other aspects of rheumatoid arthritis. One is related to extra-articular manifestations and one to disease severity now compared to 1978 indicating that although demographic data are very similar patients are doing better than two decades ago. Similar data have previously been published by Heikki Isomäki in Finland. Yeva Lindroth working full time in private practice has defended her thesis on patient education in rheumatoid arthritis and osteoarthritis under controlled conditions. She concludes that certain effects are transient and others more lasting, but that patient education well defends its place in the armamentarium of a comprehensive arthritis programme.

These were just some examples of research activities by members of *The Swedish Society for Rheumatology*, which celebrated its 50th birthday in 1996 at Södersjukhuset in Stockholm, the site where it actually was founded. The festivities were orchestrated by associate professor Ingiäld Hafström shortly before her move of the unit to Huddinge hospital. A book was published by the Society and edited by Dr Ido Leden and associate professor Ola Nived. Unfortunately, it was decided to use the Swedish language in this publication [2].

Swedish rheumatology is in the process of standardizing the initial assessment and therapy of patients with rheumatoid arthritis and entering data into a common computer program. The annual autumn meetings in Stockholm will continue but will move to Gothenburg every second year starting in 1998. In addition spring meetings will be held, next time in April of 1998 in Lund, where the author of this letter will soon add the prefix of emeritus to his title, although he would prefer the Italian word 'senator'.

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LETTERS TO THE EDITOR

HLA-B51 Negative Monozygotic Twins Discordant for Behçet's Disease

SIR—Behçet's disease (BD) is a multisystem inflammatory vasculitic disorder of unknown aetiology. Both genetic and environmental factors have been claimed to be involved in the aetiopathogenesis of BD [1]. A strong association has been known with HLA-B51 and BD in countries on the Silk Route from the Mediterranean area to Japan [2]. However, this association is not prominent in those countries where BD is not prevalent, such as the UK and USA. In addition to ethnic predilection, familial aggregation also supports the genetic predisposition to BD, and this is especially manifest in families of probands carrying HLA-B51 [3–5].

Twin concordance studies are very helpful for examining the complex interactions between genetic and environmental factors. To date, only one pair of monozygotic (MZ) twins with BD has been reported [6]. These twin brothers were HLA-B51 positive, concordant for BD and developed the disease at the same age. We report herein two more pairs of MZ twins of Turkish origin. They are both HLA-B51 negative (Table I) and currently discordant for BD. DNA polymorphism analysis using a polymarker kit (AmpliType PM, Perkin Elmer, USA) confirmed with $>99.9\%$ confidence that both twin pairs are MZ and both probands fulfilled the criteria of the International Study Group for the diagnosis of BD [7].

The first proband is a 26-yr-old male. He developed oral and genital aphthous ulcerations and bilateral posterior uveitis 5 yr ago. A skin pathergy test was found to be positive. After the diagnosis of BD, he had been on colchicine, corticosteroids, azathioprine and lately cyclophosphamide. However, he suffered from recurrent attacks of uveitis and lost his vision almost totally. He had been raised together with his twin brother, but they have been working at different jobs since the age of 14. His brother has no symptoms except oral aphthous ulcerations occurring one or two times a year or less, and skin pathergy tests have given consistently negative results.

The other proband is a 45-yr-old male. He had recurrent oral and genital ulcerations and folliculitis for 2 yr. He developed Achilles tendinitis and arthritis in his left ankle 6 months ago, and the skin pathergy test was found to be positive. He responded well to colchicine and non-steroidal anti-inflammatory drugs (NSAID). He had shared the same room of their home with his twin brother until the age of 25. His twin brother has no symptoms or signs regarding BD and his skin pathergy test was consistently negative. On the other hand, they have a 49-yr-old brother who has BD and he is HLA-B51 positive (Table I). He had recurrent attacks of acute mono/oligoarthritis since the age of 28, and started to suffer from erythema nodosum and folliculitis at the age of 32. He had recurrent oral aphthous ulcerations when he was 38, and developed bilateral posterior uveitis 1 yr later. A positive skin pathergy test and radiological evidence for enthesitis in both ankles were noted at the time of diagnosis. He has been followed up to 10 yr in our unit and is doing well on treatment with colchicine, NSAID, and topical and systemic steroids when necessary. All brothers are negative for HLA-B27 and have no family history of any form of spondyloarthropathies.

Twin concordance studies are frequently being used for estimating the role of genetic factors in the pathogenesis of multifactorial diseases, although disease concordance rates in MZ twins do not show exactly the upper level of the genetic contribution to the disease [8]. The pathogenetic role of HLA-B51 in BD has yet to be elucidated. Transgenic mice studies revealed that the HLA-B51 molecule itself might be involved in the pathogenesis of BD by causing neutrophil hyperfunction [9]. However, it is possible that a neighbouring gene linked to HLA-B51 may also predispose patients to BD. Microsatellite polymorphism studies in BD patients suggest that the susceptibility gene might be located between tumour necrosis factor and HLA-B genes [10]. The absence of an association between HLA-B51 and BD in the UK or USA even for familial cases could be explained by the lack of linkage between the susceptibility locus and HLA-B51 in these ethnic groups [11, 12].

Familial aggregation in the second family implies the presence of another susceptibility gene(s). Additive effects of HLA-B51 (or a gene linked to it) and a second

TABLE I
HLA antigens of MZ twins determined by standard serological methods

	HLA-A	HLA-B	HLA-C	HLA-DR	HLA-DQ
MZ twins-1	(A10) A26, A30	(B17) B57, B35, Bw4, Bw6	Cw3, Cw6	(DR6) DR13, DR7, DR52, DR53	
MZ twins-2	A3	B18, Bw6	Cw7	(DR6) DR14, DR52	DQ1
Elder brother	A1, A11	(B5) B51, B8, Bw4, Bw6	Cw1, Cw7	DR3, (DR6) DR14, DR52	DQ1, DQ2

gene may be responsible for a relatively severe disease course with eye involvement in the elder brother, but much stronger environmental factors might be required for the development of BD in individuals carrying only the second susceptibility gene.

In conclusion, a discordant disease course up to 5 yr in these HLA-B51-negative MZ twins further supports the important role of HLA-B51 or a non-HLA gene very close to this locus in the genetic predisposition to BD. Larger series of MZ and dizygotic twins, and investigation of the multicase families, are needed to clarify the genetic component in the pathogenesis of BD.

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Autoantibodies Against Cardiolipin and Endothelial Cells in Takayasu's Arteritis: Prevalence and Isotype Distribution

Sir—Takayasu's arteritis (TA) is a large-vessel vasculitis, with a predilection for the aortic arch and its branches. The aetiology of TA is not known. Some of the current clinical and laboratory data point towards an immune basis for the disease [1–3]. In view of the potential role of antibodies to cardiolipin (aCL) and endothelial cells (aEC) in vascular damage, we considered it of interest to study in TA patients the prevalence and levels of IgG, IgM and IgA isotypes of these two autoantibodies.

The study group consisted of 30 TA patients presenting between 1994 and 1995 at a tertiary care hospital in Lucknow, India, and 44 age- and sex-matched healthy controls. TA was diagnosed according to the ACR criteria [4] and considered to be in an active stage if two or more of the following were present: (i) constitutional features, for which no other cause could be identified; (ii) painful arteries; (iii) elevated ESR (>30 mm/h); (iv) elevated CRP (>0.6 mg/dl). Thirteen patients had active disease. None of the patients had signs of the antiphospholipid syndrome.

IgG and IgM aCL and aEC were determined according to the method described previously [5]. For IgA aCL and aEC, the serum was used in a dilution of 1/30 and anti-human IgA alkaline phosphatase conjugated (α-chain specific, Sigma, St Louis, MO, USA) in a dilution of 1/1000. The cumulative intra- and inter-assay coefficients of variation were $<10\%$ and $<15\%$ for aCL and aEC, respectively. The cut-off limit for a positive value was taken as the mean + 2 S.D. of the control group excluding three very high values of IgG aEC and one value of IgM aEC; these cases were, however, taken as positive.

The standardized normal deviate and Fisher exact test were used for comparing two proportions. The Mann–Whitney U-test was used for comparison of levels of antibodies between patient and controls.

Significantly higher levels of all three isotypes of aCL ($P < 0.01$) and of IgG ($P < 0.01$) and IgM ($P < 0.05$) isotypes of aEC were observed in patients in comparison to those in controls (Fig. 1). The prevalence of one or more isotypes of aCL was 53.3% and of aEC 36.7% in patients in comparison to 9.1% ($P < 0.01$) and 13.6% ($P < 0.05$), respectively, in controls (Table I). Furthermore, in patients with active disease, the prevalence of aCL and aEC was 84.6 and 61.5%, respectively, in comparison to 29.4% ($P < 0.05$) and 17.6% ($P < 0.05$), respectively, in those with inactive disease. aCL and aEC co-occurred in 33% of patients.

Although aCL have an established association with recurrent venous and/or arterial thromboses [6], there are now reports of aCL in inflammatory vascular diseases, as well as one on IgG aCL in TA [7–9]. Our

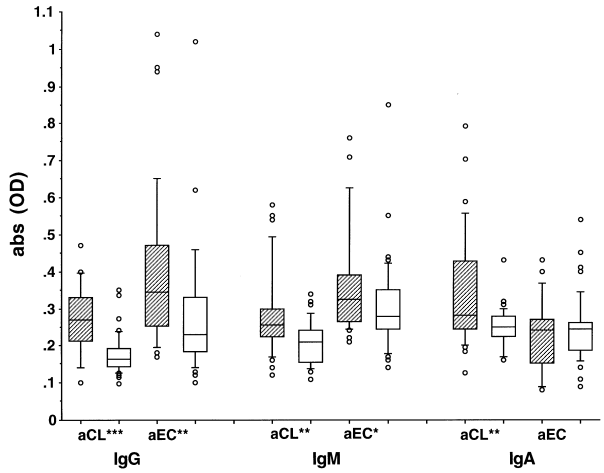

FIG. 1.—Box plots showing the levels of the three isotypes of aCL and aEC in patients (▨) and in controls (□). The levels are expressed as optical density (OD) values. The lower, mid and upper horizontal lines of the boxes represent 25th, 50th and 75th percentiles, respectively; the vertical lines extend from the 10th to the 90th percentile. $*P < 0.05$; $**P < 0.01$; $***P < 0.001$.

observation of an association of aCL with the activity of the vasculitic disease lends further support to a close relationship of these antibodies with vascular inflammation. The unusually high prevalence (33.3%) of IgA aCL observed by us may indicate that the triggering event for the formation of these antibodies is at the mucosal level, possibly a gastrointestinal or a respiratory tract infection.

The association of aEC with disease activity may also suggest their pathogenic involvement in TA. There is only one report available on aEC in TA in which IgG aEC was studied and observed to be elevated in 94% of TA patients [10]. The disease activity status of the patients in this study was not mentioned and the very high prevalence of aEC in comparison to that observed by us could be because most of their patients may have had active disease.

TABLE I
Prevalence of antibodies to cardiolipin and endothelial cells in Takayasu's arteritis patients and in healthy controls

	Patients		Controls		
	n	%	*n*	%	*P*
aCL					
IgG	10	(33.3)	2	(4.5)	< 0.01
IgM	6	(20.0)	1	(2.3)	< 0.05
IgA	10	(33.3)	1	(2.3)	< 0.01
aEC					
IgG	10	(33.3)	5	(11.7)	< 0.05
IgM	6	(18.8)	2	(4.5)	< 0.05
IgA	1	(3.3)	3	(6.8)	NS

aCL, antibodies to cardiolipin; aEC, antibodies to endothelial cells.
NS, not significant ($P > 0.05$).

In summary, we have observed aCL and aEC in a significant proportion of TA patients with a correlation with disease activity. Further studies on the mechanisms of action and antigenic targets of the autoantibodies may shed light on their pathogenic involvement in TA.

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Von Willebrand Factor Antigen and Angiotensin Converting Enzyme Levels in Takayasu Arteritis

SIR—The assessment of disease activity in Takayasu arteritis (TA) is hampered by the lack of reliable criteria and laboratory markers. Although changes in

TABLE I
Comparison of the findings among the active and inactive TA patients and the healthy and diseased controls

	Active TA patients $n = 7$	Inactive TA patients $n = 9$	PSS patients $n = 6$	Healthy controls $n = 14$	P*
ESR	83 (65–124)	27 (13–49)	51.5 (12–65)	6.5 (3–20)	0.00001
CRP	1.5 (0.7–4)	0.5 (0.5–1.8)	1.3 (0.5–3.5)	0.5 (0.5–1)	0.0008
F VIII R Ag	62 (17–150)	52 (17–83)	110 (83–154)	53 (30–140)	0.0073
ACE	14 (8–52)	13.2 (8–32)	23 (10–58)	19.8 (5.8–30.5)	0.45

Values are medians and parentheses include minimum and maximum values.
*Statistical significance: $P < 0.008$.

the erythrocyte sedimentation rate (ESR) seem to be useful in monitoring the disease activity, the finding of histological active disease despite normal ESR levels [1] makes the search for additional markers mandatory.

Since elevated levels of von Willebrand factor antigen (vWF Ag) and angiotensin converting enzyme (ACE) are considered as possible markers of endothelial damage [2, 3], we tested their levels in patients with TA. Sixteen patients (15 female, one male; mean age 35.8 $\pm$ 13.2 S.D. years) fulfilling the ACR criteria for TA [4] were registered in our clinic between 1984 and 1995. These patients were called back for the determination of ESR (Westergren), C-reactive protein (CRP) (Behringswerke AG, Marburg, Germany), vWF Ag [Malakit, Biolab products, Wavre (Limal)] and ACE levels (Sigma, Daisenhof, Germany). Their median disease duration was 9.5 months (interquartile range 1–36 months), 81% (13/16) were under treatment during the study (azathioprine alone: 3; steroid alone: 2; azathioprine and steroid: 6; cyclophosphamide and steroid: 2) and seven had active disease using the criteria suggested by Kerr *et al*. [1]. Six age- and sex-matched patients with progressive systemic sclerosis (PSS) and 14 healthy individuals were the controls. Kruskal–Wallis one-way ANOVA test was used for the analysis.

ESR and CRP were significantly elevated in TA patients with active disease and PSS patients compared to TA patients with inactive disease and healthy controls. The levels of vWF Ag were found to be increased only in PSS patients, and they did not differ between TA patients and healthy controls. ACE levels also did not differ between the groups (Table I).

In this study, despite higher levels of ESR and CRP, we failed to show an increase in vWF Ag and ACE among active TA patients. Until now, vWF Ag levels have been tested in two studies. Richardson *et al*. [5], defining active disease as an extension of angiographic findings, found elevation of vWF Ag, but McRorie *et al*. [3], without explaining the method used for assessing disease activity, did not.

Active immunosuppressive treatment and long-standing disease could explain our results. Furthermore, endothelial damage might not be prominent in the late occlusive phase of TA. Not forgetting the need for prospective studies, we conclude that vWF Ag and ACE are not helpful in assessing disease activity and monitoring the treatment of TA.

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Madelung's Deformity from a Rheumatologist's Point of View

Sir—We would like to call attention to an uncommon but confusing deformity in a clinical rheumatology setting. Madelung's deformity is described as a premature fusion of the distal growth plate in the radius with overgrowth of the ulna, decreased radial length and dorsal subluxation of the distal ulna resulting in a non-inflammatory deformity of the wrist [1–3]. This condition is idiopathic, but may be associated with other hereditary or congenital disorders such as dyschondrosteosis [4–7]. Madelung's deformity is more prevalent among women and could be influenced by *in utero* sex hormones or by the X chromosome [8].

We describe the case of an 11-yr-old girl who was sent to us with arthralgia and deformity in both wrists

of 2 yr duration. She was the fifth child born to unrelated healthy parents aged 27 (mother) and 34 yr at the time of conception. Her two brothers and two sisters were alive and healthy. There was no familial history of a similar disorder. History of arthralgia or arthritis was negative. On physical examination, we found that the distal aspects of the right and left ulnae were prominent in a lateral view; dorsiflexion and palmar flexion of both wrists were limited, without evidence of synovitis. In the lower extremities, her knees showed a tendency to valgus deformity. No other significant abnormalities were found. The radiological findings showed distal radial epiphysis triangular and deviated in an ulnar and volar direction. The ulna was dorsally subluxed in relationship with the radius. The carpal bones were wedged into a triangular configuration between the obliquely radial head and the displaced ulnae (Fig. 1). Laboratory and serological data, including blood cell count, glucose, urea, creatinine, coagulation studies, urinalysis, liver function tests, rheumatoid factor, erythrocyte sedimentation rate (Westergren), VDRL test, serum complement levels, antinuclear antibodies and antibodies to nDNA detected by indirect immunofluorescence (using HEp-2 cells and *Crithidia luciliae*, respectively), were all normal or negative. She received acetaminophen as the only medication, providing relief of her arthralgia symptoms.

Clinical manifestations of primary Madelung's deformity typically become evident during adolescence. It tends to be bilateral and initially is usually painless. The findings seen in this condition could be confusing in general clinical practice due to more advanced deformity which may mimick those seen in advanced rheumatic conditions like juvenile chronic arthritis (JCA). However, there are differences, since overall Madelung's deformity is not an inflammatory disease. In our case, the patient was referred to us with the diagnosis of JCA. Madelung's deformity was suspected on the basis of anatomical deformity at both wrists without arthritis and confirmed by the radiological findings. Genetic counselling was provided as a sporadic non-familial case on the basis of a negative family history, unaffected parents and siblings. A new autosomal dominant mutation cannot be excluded.

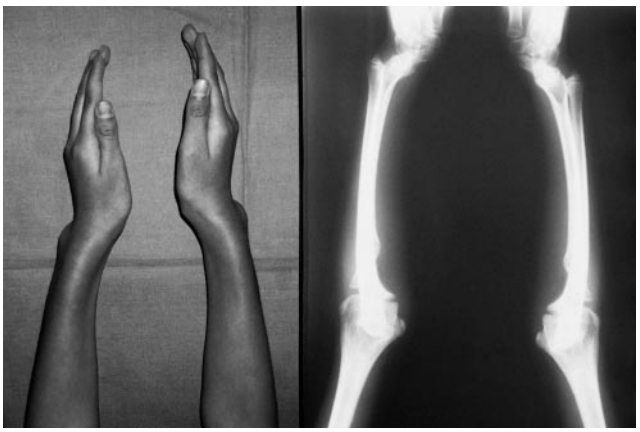

FIG. 1.—Lateral radiograph of the forearm in which the plane of the distal articular radial surface has exaggerated volar and ulnar tilt. The hand and carpus are subluxed ventrally.

Appropriate identification of Madelung's deformity is important to avoid an incorrect diagnosis of deformity due to acute injury or to an inflammatory rheumatic disease.

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Shared DMARD Monitoring

SIR—We have previously presented an audit of disease-modifying anti-rheumatic drug (DMARD) monitoring to illustrate the shared care of drug monitoring between hospital and general practice [1]. We identified three key elements for such a scheme to work: patient information leaflets, general practice information leaflets and shared monitoring cards carried by the patient. As a result of the audit, certain changes were implemented, notably the production of customized monitoring cards, the distribution of comprehensive general practice information packs, a telephone help line for problems and recording, by pathology laboratory staff, of blood test results in patient monitoring cards.

Two years after these changes were made, we have re-audited our DMARD monitoring on a sample of 100 consecutive patients (taking 110 DMARDs) attending the rheumatology out-patient department. The results (see Table I: only patients taking D-penicillamine, sulphasalazine, gold injections,

TABLE I
Comparison of 1993 and 1995 audits

	D-penicillamine		Sulphasalazine		Sodium aurothiomalate		Azathioprine		Methotrexate	
	1993	1995	1993	1995	1993	1995	1993	1995	1993	1995
N	41	4	112	53	34	5	17	10	29	20
Age (mean, yr)	55.2	59.0	59.9	58.0	55.4	58.6	54.9	58.1	58.1	53.5
Duration of disease (mean, yr)	15.4	28.5	8.8	9.7	10.4	16.3	10.2	4.7	14.2	11.8
Duration of use (mean, months)	64.9	69.8	18.1	38.7	27.5	84.4	7.2	18.9	8.7	29.2
Adequate patient monitoring record (number with)	4	2	8	39	30	5	3	10	13	15
Information sheet supplied (number with)	23	0	81	42	28	2	6	10	29	19
GP supplied with copy of protocol (number sent)	24	1	84	42	28	4	14	9	28	19
Monitoring protocol followed (%)	28 (68)	4 (100)	75 (67)	39 (74)	9 (26)	5 (100)	12 (71)	9 (90)	27 (93)	19 (95)
Mean protocol monitoring score (maximum = 4.0)	1.9	1.5	2.2	3.3	2.8	3.5	2.1	3.9	3.3	3.7

azathioprine and methotrexate included) indicate an improvement in the percentage of patients in whom the defined monitoring protocol was followed (overall 65% in 1993; 83% in 1995). As expected, more patients were found to be carrying an up-to-date monitoring card (22% in 1993; 77% in 1995) and substantial improvements in the mean monitoring score (as defined in our previous paper) were found. The main reason for protocol failure was, as before, lack of appropriate blood tests.

Although the results are not ideal, we believe that shared monitoring schemes can work providing the key elements of information, communication and patient-held cards are provided. As the responsibility for safety lies with the prescriber, it may improve monitoring schedules if the general practitioner insists on the production of an up-to-date monitoring card before a repeat prescription for the DMARD is issued.

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