nodosum leprosum and lupus profundus panniculitis, immunologic mediated conditions responding well to thalidomide. There has been so far only one other report of thalidomide use in WCD. 24 years ago a favourable response in a 23 y.o. Female from Malaysia was reported. This patient had a remission, which was ongoing despite stopping of all medications after treatment with thalidomide of only 13 weeks. The question of a possible spontaneous remission asked by the describing authors remains unanswered. To the contrary, in the here described case repeated reduction of thalidomide below 100 mg/dl caused mild recurrence of sympt., responding immediately to dose increase to 100 mg.

Thalidomide's anti-TNF α modulatory and anti-angiogenic effects have recently been rediscovered also for treatment of other autoimmune diseases. Superior tolerability and side-effect profile (compared to corticosteroids and immunosuppressives) make thalidomide a first line treatment consideration for chronic WCD in patients without childbearing potential.

THU0219 RETROSPECTIVE EVALUATION OF 180 PATIENTS WHO WERE DIAGNOSED AS UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE

E Doganavsargil, F Oksel, G Keser, V Inal, K Aksu. *Rheumatology, Ege University Hospital, Izmir, Turkey*

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Background

Objectives To review the outcome and diagnosis of patients whose initial clinical and laboratory findings were suggestive of a well established and undifferentiated connective tissue disease (uCTD), without fulfilling the diagnostic criteria of any connective tissue disease.

Methods Medical records of patients being followed up by Ege University rheumatology department between 1990 and 1999 were retrospectively screened. The data of 478 patients, constituting 18% of the total, whose initial diagnosis was uCTD, were further investigated. Patients with missing data were omitted and only 180 patients having reliable and complete initial clinical and laboratory data with regular controls were included.

Results The mean ages of 180 patients (F/M:173/7) at the time of initial clinical symptoms and at the time of diagnosis of uCTD were 35 ± 12 years and 38 ± 12 years, respectively. The distribution of initial symptoms and clinical findings were as follows: arthralgia/arthritis 97.2%, Raynaud phenomenon (RP) 30.5%, mucocutaneous findings 68.2%, symptoms of keratoconjunctivitis sicca 14.4%. It was notable that none of the patients had presented with initial symptoms or clinical finding reflecting direct involvement of a specific organ.

In the final evaluation, 46 out of 180 patients (25.5%) were found to evolve into a definite clinical diagnosis: Sjögren's syndrome 21 (11.6%), systemic lupus erythematosus 8 (4.4%), rheumatoid arthritis 8 (4.4%), CREST syndrome 7 (3.8%), diffuse systemic sclerosis 1 and seronegative polyarthritis 1. In 5 patients a nonrheumatologic diagnosis was made, while in 11 patients fibromyalgia was diagnosed. The diagnosis of 118 (65%) patients (F/M:115/3) remained unchanged.

The evaluation of initial clinical and laboratory parameters with respect to their contribution to final diagnosis revealed that, presence of Raynaud phenomenon and anti-centromeric antibodies were highly suggestive of later diagnosis of limited Ss. On the other hand, anti-dsDNA antibodies, leucopenia and

thrombocytopenia, which are important parameters in well established SLE, were initially unproductive of later SLE development in patients whose initial diagnosis was uCTD.

Conclusion In most of the patients whose initial diagnosis was uCTD, the disease tends to remain undifferentiated.

THU0220 AN ESTIMATION OF THE PREVALENCE OF HYPERMOBILITY

¹M Gumà, ²J Roca, ¹S Holgado, ¹M Valls-Roc, ¹E Casado, ³J Forcada, ¹JC Duro, ¹A Olive, ¹X Tena. ¹Rheumatology, ²Epidemiology, ³GP, Hospital Universitari Germans Trias I Pujol, Badalona, Spain

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Background Hypermobility is associated with several rheumatic manifestations. The association with SLE is controversial.

Objectives To determine the prevalence of hypermobility in two different populations: a general practitioner (GP) office and out-patients clinic of a referral rheumatology hospital.

Methods 280 patients (193 females, 87 males); mean age 44.7 ± 11.2 were consecutively reviewed in a GP office. 370 patients (233 females, 84 males); mean age 50.5 ± 16.8 were consecutively visited at the out-patients rheumatology clinic. Hypermobility was evaluated following the criteria of Carter-Beighton (hypermobility > 4 points).

Results Hypermobility was present in 42/280 (15%) at the GP office and in 81/317 (25%) in the out patients rheumatology clinic.

Abstract THU0220 Table 1

	General practitioner		Hospital	
Hypermobility	YES	NO	YES	NO
Age	28 ± 11.7	47 ± 18.3	38.9 ± 14.2	54 ± 15.9
Sex (F/M)	5.8/1	1/1.2	3.7/1	1/1.3
Criteria	5.1 ± 1	0.6 ± 1	5.6 ± 1.6	0.5 ± 0.9

Conclusion The prevalence of hypermobility at the GP office and a rheumatology clinic is high. Furthermore it is close to the prevalence of hypermobility in the general population.

THU0221 OSTEONECROSIS AND HIV INFECTION: REPORT OF ELEVEN CASES

¹M Valls-Roc, ¹V Ortiz-Santamaria, ¹A Olive, ²M Cervantes, ²J Gratacos, ³G Sirera, ³C Tural, ³E Negro, ¹X Tena, ³B Clotet. ¹Rheumatology Service, Hospital Universitari Germans Trias I Pujol, Badalona, Spain; ²Rheumatology Service, Consorci Sanitari Parc Taulí, Sabadell, Spain; ³HIV Unit, Hospital Universitari Germans Trias I Pujol, Badalona, Spain

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Background A vascular necrosis (AVN) has been described in the setting of HIV infection. No epidemiological studies to determine the higher incidence of AVN in HIV+ patients are available. Protease inhibitor containing regimens (PI-CR) have been considered an additional risk factor for developing AVN.

Objectives To report the clinical findings of 11 HIV + patients with AVN.

Methods Retrospective review of clinical and radiological charts of 11 patients diagnosed of AVN.

Results Eleven patients were diagnosed of having AVN: Eight male and three female. The mean age was 27.6 years (23–32). The risk factors for HIV infection was as follows: 8 IDU, 3 HMX. Nine patients had had at least one opportunistic infection with a mean time of 30.5 months before the diagnosis of the AVN (12–108). The mean CD4⁺ and pVL at AVN were 328.9 cel/mm³ (73–900) and 14.964 copies/ml (6 of them had undetectable levels: <200 copies/ml) respectively. Mean CD4⁺ nadir was 53 cel/mm³. The mean time on PI-CR before the AVN was 19.4 months (2 patients were naïve for PIs before AVN). MRI showed a bilateral femoral head involvement in 9 patients, 2 of them with additional humeral and knee involvement. Other risk factor related to the development of AVN were evaluated: 6 patients had received corticosteroids prior to the AVN, hyperlipidaemia secondary to PI-CR was reported only in 2 patients. Six months before the AVN, salmonella typhimurium bacteriemia was documented in 2 patients.

Conclusion Despite the higher incidence of AVN reported in HAART era, epidemiological studies should be conducted in order to establish the role of PIs in its aetiology. Other risk factor such as corticosteroids use related to AVN, should be assessed in HIV+ patients with AVN.

THU0222 TRANSIENT BONE MARROW OEDEMA AND/OR TRANSIENT OSTEOPOROSIS OF THE HIP ARE ASSOCIATED WITH GENERALISED DIMINUTION IN BONE MINERAL DENSITY: ROLE OF PRE-EXISTING BONE FRAGILITY ?

¹T Pham, ¹V Lègré, ²P Champsaur, ²C Chagnaud, ¹P Lafforgue. ¹Department of Rheumatology; ²Department of Radiology, CHU La Timone, Marseille, France

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Background Transient osteoporosis of the hip (TOH) and transient bone marrow oedema of the hip (TBMEH) are uncommon, closely associated and/or similar conditions, whose pathophysiology remains obscure. Several facts suggest that occult microfractures of the femoral head could be the cause of such entities.

Objectives We conducted a prospective assessment of bone status in skeletal sites other than the affected hips to test the hypothesis of pre-existing underlying generalised bone fragility.

Methods 13 consecutive patients with typical signs of TBMEH at MRI were enrolled, with careful exclusion of cases presenting with any sign consistent with either osteonecrosis or stress fractures of the femoral head. Five patients also had radiological signs of TOH. All had prospective bone status assessment including search for risk factors of bone fragility or history of fracture, laboratory tests, spine radiographs and lumbar and femoral neck (of the unaffected hip) by DEXA.

Results There were 8 women (6 pregnant) and 5 men, aged 40 ± 11 years. Four had possible risk factors for osteoporosis, and only 1 had previous insufficiency fracture (rib). Lumbar BMD measurements by DEXA found 1 patient with normal BMD (T-score between 0 and 1SD), 9 patients had a T-score between 1 and 2.5 SD and 2 had T-score less than 2.5 SD. The BMD values were about 15% less than the age/sex matched reference population (mean Z-score 1.4 ± 0.64 SD). BMD values obtained in the unaffected hip in 6 cases confirmed these results. Separate

analysis of males/females and pregnant/other patients yielded the same results.

Conclusion Most patients with TBMEH and/or TOH have a generalised decrease in BMD. These results put together with data from the literature suggest that a pre-existing bone fragility is a predisposing factor to TBMEH and/or TOH.

THU0223 TREATMENT OF A PATIENT WITH NONCLASSIFIED HLA B27 ASSOCIATED DESTRUCTIVE OLIGOARTHRITIS WITH ANTI TNF: CLINICAL OUTCOME AND EFFECTS ON NF-KAPPA B ACTIVITY IN PBMC

T Geiler, R Voll, HM Lorenz, C Dechant, B Manger, JR Kalden. Department of Medicine III, University of Erlangen-Nuremberg, Erlangen, Germany

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Background We report the case of a 38 years old HLA B27 positive male with a 11 year history of a destructive nonclassified oligoarthritis. While the first 6 years of the disease have been dominated by a relapsing nondestructive synovitis of the right knee joint and a asymptomatic sacroileitis treated with NSAID's alone the disease began to spread in a destructive manner to the right wrist and foot (MTP I-V, PIP I-V, DIP I-V). Additionally, a painful tenosynovitis of the flexor and extensor tendons of both feet occurred. Despite intensifying the pharmacological treatment by high doses of corticosteroids (up to 100 mg/d) and methotrexate (25 mg/week) for 9 month the disease showed significant clinical and radiological progress. No reduction of the constantly elevated ESR and CRP could be achieved. By initiating a therapy with anti TNF (Remicade™) at a dose of 5 mg/kg body weight all clinical symptoms disappeared completely within 8 h after the first infusion of the antibody. ESR and CRP normalised within 6 days. The patient stayed completely free of symptoms since, side effects have not been detected by now. Repeated infusions after 2, 4, 6 and 14 weeks did not further improve the therapeutic benefit but were given to sustain the therapeutic effect.

Objectives Since TNF is known as a strong activator of NF-κB which is strongly involved in the production of proinflammatory cytokines and matrix metalloproteinases we looked for the activity of this transcription factor in PBMC's of this patient by electro mobility shift assays (EMSA).

Methods Cells were obtained immediately before the infusions were started and 1 h and 24 h after end of infusions. Cell preparations and EMSA were done according standard protocols.

Results There was substantial NF-κB activity in PBMC's before first antibody treatment. Anti TNF treatment decreased NF-κB activity detectable already 1 h after first infusion. The NF-κB activity was further inhibited 24 h after first infusion and remained low during the whole treatment period in accordance to the persisting clinical improvement.

Conclusion In the treatment of patients with progressive non-classified HLA B27 oligoarthritis resistant to classical treatment with NSAID's, corticosteroids and DMARD's the use of TNF blocking agents should be considered. As in other destructive inflammatory joint diseases NF-κB might play a crucial role in the pathogenesis of joint destruction in HLA B27 associated arthritides. The clinical improvement appears to be associated with a persistent and marked inhibition of NF-κappa B.