877 **Complement deficiencies in patients affected by Meningitis caused by *N. meningitidis*.** AS Grumach MD, A Campeas MD, CL Diogo BS, MC Raymundo MD, LC Oliveira BS, MSS Carneiro MD, M Kirschfink PhD, São Paulo, Brazil

Terminal components (C5-9) of classical pathway of Complement System and properdin deficiencies (PD) have been described in patients affected by meningitis. This study evaluated 90 patients (54 males and 46 females) with meningitis caused by *Neisseriae meninigitidis*. The bacterial identification was done through bacterioscopy (negative Gram *Diplococcus*) and/or latex agglutination and/or LCR culture. Patients mean age was 61 months (1 month-23 years). In 37 cases, *N. meningitidis* types were: B (n=17), C(n=18) and W135 (n=2). Initial Complement evaluation was performed with the following tests: Hemolytic assays (CH50 and APH50) and properdin levels (ELISA). Specific complement components were analysed for low CH50 and APH50 values. Complete absence of CH50 (C2 and C6 deficiency) and PD were detected in 2 and 6 patients, respectively. The detection of complement deficiencies predisposing to meningitis is important in order to control this infection earlier. Regarding properdin deficiency, the *N. meningitidis* vaccine is capable of preventing other infections.

878 **A novel familial C2 abnormality.** JS Seggev MD*, P Giclas PhD**, Las Vegas, NV*, & Denver, CO**

C2 deficiency presents phenotypically reduced serum (0-10% of normal) concentration and absent or very low hemolytic activity. Heterozygous patients have levels and function that are about half the normal values. We herein describe a family with low C2 function, in spite of normal C2 protein concentration by radial immunodiffusion. The propositus, a 9 year-old girl of mixed Chinese and American extraction, suffered staphylococcal arthritis of right hip joint at age 2y; pneumonia at age 4; developed significant hepatosplenomegaly at age 8; Asthma and allergic rhinitis since the age of 2. Immunological evaluation revealed normal B lymphocyte function; absent skin DTH reactions; normal T and subset lymphocyte numbers and in vitro proliferation to mitogens and antigens; normal NBT reduction. HLA phenotype is A1, A2, B13, B44, Cw5, Cw6, DR1, DR15. CH50 was consistently reduced to less than 50% of normal, while alternative pathway AH50 was normal. C1, C2 and C4 levels were normal, but C2 function was 27% below the lower limit of the normal range. C2 function of 3 siblings, 2 maternal nephews and her maternal grandmother had C2 function ranging from 50% to 5% below the lower limit of normal, her mother was at the lower limit, and her father, uncle, and maternal grandfather had normal C2 function. The C2 levels were normal to high for all family members. Studies to characterize the abnormal C2 protein are under way. No reports of a dysfunctional C2 protein have been found on a Medline search. The relation-ship of this abnormality to our propositus' clinical presentation is unclear. Because the C2 gene is located in the MHC region on chromosome 6 , the C2 abnormality may reflect another subtle immune defect.

879 **Properdin Deficiency: Rare Presentation with Meningococcal Osteomyelitis and Arthritis.** M Rottem MD, E Shiloach MD, M Schlezinger MD, Afula, Zrifin and Ashkelon, Israel.

Properdin deficiency is an X-linked rare form of immunodeficiency of the complement system. It often presents with fatal meningococcal meningitis. We have recently diagnosed and treated two patients with properdin deficiency. The first was a 13 years old boy who presented with meningococcal osteomylitis and meningococcal purpuric lesions as demonstrated by skin biopsy. Family history was significant for a 14 years old brother and a maternal uncle who had died two years earlier of fulminant and meningococcal meningitis. Complement studies revealed reduced AP50 levels and properdin levels in the propositus and his two living brothers, but normal levels in their parents (57, 63, 79, 144, and 159 U/ml, respectively, and 0.27, 0.19, 0.27, 1.44, and 1.65 U/ml, respectively). AP50 and properdin levels were normal in 3 sons and daughter of the deceased maternal uncle. Parenteral antibiotic treatment led to complete recovery. The second patient was a 20 years old male who presented with meningococcal arthritis. His family history was uneventful. Complement studies revealed low AP50 and markedly reduced properdin level. He also was treated successfully with parenteral antibiotics and recovered. In conclusion, these two cases demonstrate that meningococcal infections in some patients with properdin deficiency may present with meningococcal osteomyelitis or arthritis. Further studies are necessary to determine the nature of this predilection. Treated promptly, meningococcal infections in such cases have favorable outcome.

880 **Immunomodulatory effects of levamisole in chronic fatigue syndrome**

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Chronic fatigue syndrome (CFS) may be associated with immunologic abnormalities. We describe a patient with CFS and coexisting lymphopenia and cellular immunodeficiency who responded to treatment with levamisole. A 32 y.o. female developed CFS after a blood transfusion for post partum bleeding 4 years ago and since then she reports low grade fevers, insomnia, myalgia, and severe lethargy. She had intermittent oral candidiasis which was successfully treated with antifungals. Oral lesions consistent with Herpes simplex were observed but a trial of acyclovir failed to cause significant improvement. Laboratory evaluation was negative for active CMV, EBV or HIV infections as were seroassays for hepatitis B, lyme disease, and autoimmune disorders. Throat cultures were negative. Muscle enzymes and thyroid studies, IgG, IgM, IgA, IgG subclasses, pneumococcal, tetanus, and viral specific antibody titers were normal. Flow cytometry showed normal numbers of T and B lymphocytes but the CD4 count was in the low normal range. Functional immunoassays revealed diminished proliferative responses to mitogens and antigens. A trial of the immunopotentiator, levamisole, at a dose of 50 mg 3 times daily for 3 days every 2 weeks was given for 3 months. At the end of this period the patient experienced considerable improvement of subjective symptoms, including increased stamina. More objectively, this was accompanied by significant changes in immunologic parameters, including an increase in CD4 counts and a major increase in lymphocyte proliferative responses to mitogens and antigens. The patient continues to do well for the past 6 months without specific therapy. This case suggests that the immunopotentiator, levamisole, may be useful in the management of CFS associated with objective evidence of coexisting cellular immunodeficiency.