age of ten years. The following years symptoms worsened and she developed headaches, depressive thoughts with catatonic as well as obsessive-compulsive and psychotic behaviour with multiple suicidal attempatients. Laboratory investigations revealed leuco- and thrombopenia, as well as a positive ANF, with anti-ENA RNP-a, RNP 70-k and RNP-c, suggestive for an auto-immune disorder. MRI of the brain was normal, but EEG was abnormal and the skin biopsy revealed IgG-deposition along the basal membrane. Initial treatment with corticoids and hydroxychloroquine resulted in improvement. However, after three months symptoms worsened and were resistant to subsequent treatment (Corticoids, low dose cyclophosphamide, plasmapheresis and anti- CD20 monoclonal antibodies). Finally, an ASCT according to the EBMT- ASTIL guidelines was performed. PBSC harvest was done after Cyclophosphamide (2X2g/m).The conditioning regimen consisted of cyclophosphamide 200 mg/kg and ATG 7.5 mg/kg. There were no severe problems with the conditioning regimen. The ASCT resulted in a spectacular improvement of the neuropsychiatric disabilities of this patient.

Discussion
Patients with NPSLE who are refractory to conventional treatment can achieve clinical remission after ASCT. This improvement is due to immunological changes that can not be achieved with other treatments. Patients should be considered for ASCT if a) they are resistant to first line treatment b) there is a high mortality risk due to de NPSLE, in this case suicidal attempatients c) ASCT should be performed before irreversible organ damage has occurred.

P.73 The immunomodulating peptide thymosin alpha 1 has no effects on multiple myeloma evolution and on immune reconstitution

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Background
Thymosin alpha 1 (Ta1) is a thymic peptide with known immuno-modulating properties, including increase of immune reconstitution after stem cell transplantation (SCT), enhancement of anti-tumour responses and direct effects on cancer cells (up-regulation of MHC-I expression, decrease of proliferation). We thus wanted to study the effects of Ta1 on murine models of multiple myeloma (MM) and immune reconstitution after SCT.

Methods and results
We first studied the *in vitro* effects of Ta1 on two murine MM cell lines, MOPC315.BM (Balb/c background) and 5TGM1 cells (C57Bl/KalwRij background). No significant effect of Ta1 has been observed on MHC-I/II up-regulation (flow cytometry), viability (MTT tests) or proliferation (3H-thymidin incorporation) of these cells. Since anti-tumour responses are complex and difficult to transpose *in vitro*, we studied the *in vivo* effects of Ta1 in the MM models. Mice were intravenously injected with 2.5x10^5 MM cells and treated daily by subcutaneous injections with 0.4 mg/kg Ta1 or PBS. MM development was evaluated by bone marrow and spleen infiltration at sacrifice, and paraprotein quantitation. No significant, recurrent effect of Ta1 was seen on MM development. Moreover, no significant effect of Ta1 could be observed on solid tumour growth (subcutaneous injection of MOPC315.BM cells). Finally, we studied the effects of Ta1 on immune reconstitution in a humanized murine model using immunodeficient NSG mice, transplanted with 5x10^5 human hematopoietic stem cells (AC133). Immune cell reconstitution was studied by flow cytometry on blood samples and at sacrifice (105 days post-transplantation). No significant effect of Ta1 could be seen on immune reconstitution in this model.

Conclusions
No biological effects of Ta1 could be observed on MM development or immune reconstitution in the proposed murine models, in contradiction with the immunomodulatory properties attributed to this peptide in the literature.

P.74 Rothmund-Thomson Syndrome: Immuno-Osseous challenges

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Background
Rothmund-Thomson (RTS) syndrome is a rare autosomal recessively inherited genodermatosis. It is characterized by poikiloderma, small stature, skeletal and dental abnormalities, cataract, and an increased risk of cancer. The syndrome is caused by mutations in RECQL4 at 8q24.

Aims
To describe the osseous and immunologic features of three patients with genetically confirmed RTS.

Methods
Immunological investigation, x-ray imaging and bone densitometry were performed at time of the first visit to the combined rheumatology-immunology clinic.

Results
All patients had characteristic poikiloderma as well as thumb anomalies. They were born dysmaturely and presented with failure to thrive. Age at genetic diagnosis was 5y, 4y and 3y for P1, P2, P3. Osteopenia and abnormal metaphyseal trabeculation of bones were striking on the initial skeletal survey in all patients. Z-scores on DXA scan were -0.1, -1.1 and -1.2 for P1, P2, P3 respectively at presentation. The presentation in P2 was dramatic with six fractures in upper and lower extremities and subluxation of both radii.
All patients were suffering from recurrent chest infections. P1 had granulomatous skin inflammation following primo VZV infection. All patients have low switched memory B cells for age, P1 has IgG2 deficiency. P1 and P3 have IgM deficiency. P1 and P3 have specific polysaccharide antibody deficiency. Results are pending for P2. All receive prophylactic antibiotics. P1 is treated with subcutaneous immunoglobulin substitution.

Conclusion
RTS is a genodermatosis with variable clinical presentation and course. Our observation of severe bone abnormalities and associated immunodeficiency merits attention for optimal management of these patients.

P.75 Invasive Aspergillus pneumonitis as a first presentation of X-linked Chronic Granulomatous Disease (X-CGD)

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Introduction
Chronic granulomatous disease is a primary immunodeficiency disorder characterized by recurrent life-threatening bacterial and fungal infections and granuloma formation. CGD is caused by