APS, pDC and mDC produced BAFF and expressed chemokine receptors.

Conclusion pDC and mDC are differentially affected by IFNα in SLE and APS. IFNα primes pDC for enhanced IFNα production which potentiates T-cell activation by mDC, thereby sustaining the IFN signature in SLE and APS.

**PS5:92** CLARIFICATION OF THE ROLE OF DNASE 1 ON THE ONSET OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A MURINE MODEL
1E Kenny, 2U Abu Abed, 3A Kuetei, 4B Raupach, 5V Brinkmann, 6A Zychlinsky. 1Max-Planck-Institute for infection biology, dept. of Cellular Microbiology, Berlin, Germany; 2Max-Planck-Institute for infection biology, Microscopy Core Facility, Berlin, Germany; 3Charite Universitaetsmedizin Berlin Forschungszentrum Immunwissenschaft RCS, Berlin, Germany

Systemic lupus erythematosus (SLE) is a prototypical autoimmune disease resulting in multi-organ damage and a high rate of morbidity. Onset of SLE is characterised by dysregulated activation of T and B lymphocytes and the production of autoantibodies directed against nuclear components. The autoantibodies generated during the onset of SLE often recognise components released by neutrophils during NETosis, a type of cell death defined by the generation of neutrophil extracellular traps (NETs). The endonuclease DNase1 has been shown to be involved in the clearance of NET components. The sera of SLE patients contain inhibitors of DNase 1 and/or anti-NET antibodies that block the ability of DNase 1 to degrade NETs. Thus, whilst NETs are important for clearing infection they must be tightly regulated and degraded to prevent the onset of autoimmunity.

In this study we monitored the production of autoantibodies in the serum of wild type and DNase 1-deficient mice from the age of 2 to 12 months, along with proteinuria levels and the development of glomerulonephritis. We show that DNase 1-deficient mice develop a SLE-like phenotype with elevated autoantibody production and kidney damage by 12 months. This model also demonstrates the female bias in SLE as the female DNase 1-deficient mice had the highest level of kidney damage. As DNase 1 activity, B cells and aberrant NETosis are central to progression of SLE understanding their mechanisms of action are of great therapeutic interest.

**PS5:93** MARGINAL-ZONE-LIKE B CELLS DEFICIENCY REPEATEDLY DETECTED IN PERIPHERAL BLOOD AS A POSSIBLE BIOMARKER OF HYPOSPLENISIM/ASPLENIA IN SLE
1Z Hrnčíř Z, 2O Vokurkova, 3M Drahoslova, 4T Soukup, 5Tomas. 1Ind Department of Internal Medicine, Charles University Hospital, Hradec Králové, Czech Republic; 2Department of Immunology and Allergy, Charles University Hospital, Hradec Králové, Czech Republic.

Objective SLE is a disease associated with a risk of serious infections, in case of hypoplasemia/asplenia especially by encapsulated bacteria. For opsonization and phagocytosis of these agents are essential IgM natural Abs, produced only by B cells of the splenic marginal zone. Significant deficiency of marginal-zone-like B cell CD19 +CD27+IgM+B cell subpopulation absolute values x10–6/L in peripheral blood (PB) was demonstrated in a prospective, comparative, cross-over SLE study.

The data obtained demonstrated persistent character of marginal-zone-like B cells deficiency in peripheral blood, and are suggesting as possible biomarker of functional hypoplasemia/asplenia in SLE.

**REFERENCE**

Acknowledgement Supported by the research project PROGRESS Q40–15.

**PS5:94** CHARACTERISATION OF SLE B CELLS FROM PATIENTS IN REMISSION – PERSISTENT IL-10 SECRETORY DEFECT
M Siekierka-Harinek, M Schroeder, R Brinks, B Oppenorth, J Richter, S Vordenbäumen, M Schneider, G Pongratz. Rheumatology, Medical Faculty Heinrich-Heine University, Düsseldorf, Germany

Background SLE is an autoimmune disorder characterised by polyclonal Bcell activation, the production of anti-double stranded (ds) DNA autoantibodies and cytokines. Molecular and clinical studies regarding SLE often address clinically active patients and not patients in remission. This study reports on immunoglobulin, anti-dsDNA-aab and IL-10 secretory capacity of cultures of CD19 +lymphocytes from SLE patients in remission in comparison to normal donors. The aim was to evaluate whether endogenous factors (BAFF, CD40, IL4), exogenous factors (CpG-ODN-motifs, SAC) or their combinations differentially influence immunoglobulin, cytokine and anti-dsDNA-aab production in not active SLE patients vs healthy controls.

Methods Blood samples were obtained from a group of 13 SLE patients attending clinics at the rheumatology unit at the Heinrich-