vascular and 5% gastro-intestinal involvement. 37 patients (31%) were out of work with 44 patients (37%) claiming employment related benefits. With regard to individual clinical manifestations, ocular disease had a statistically significant increased risk for being out of work when assessed alone with OR 2.84 (95% CI 1.13, 7.13) but lost statistical significance when analysed in the multivariate model: OR 2.45 (95% CI 0.70, 8.60). With regard to cumulative clinical manifestations, patients with four or more clinical manifestations of BS had a statistically significant increased risk of being out of work with OR 5.57 (95% CI 1.33, 23.27) in comparison to patients with two manifestations in the multivariate model.

Conclusions: This study has highlighted the significant burden of BS on work outcomes in this UK cohort. In particular, four or more cumulative BS manifestations were independently associated with being out of work in this young cohort. Further work is required to identify whether education or intervention in the workplace can help prevent disease related job loss in BS.

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SAT0620

FURTHER EVIDENCES OF SECONDARY AMYLOIDOSIS IN ALKAPTONURIA

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Background: Alkaptonuria (AKU) is an ultra-rare inborn error of metabolism due to a deficient activity of homogentisate 1,2-dioxygenase. Patients suffer from a severe arthropathy. Evidence was provided on the presence of a secondary serum amyloid A (SAA)-based amyloidosis[¹Here a complete microscopic and ultrastructural analysis of different AKU tissues, taken from six differently aged patients, is presented.

Objectives: SAA amyloidosis is a complication in AKU[² making the detection of amyloid deposits at an early phase, important for treatment[³ We present a study of tissues from patients of different age and relevance of symptoms, providing a detailed overview of AKU amyloidosis.

Methods: Different tissues were obtained from a cohort of 6 AKU patients: 4 M (63,68,42,44 y) and 2 F (66,71 y) with different severity of symptoms. Histology and amyloid were investigated. A complete microscopic and ultrastructural analysis is presented and patient features as radiological examination, mild-to-severe degenerative changes as joint space narrowing, cartilage irregularities, sub-chondral sclerosis or peripheral osteophytes and linear intervertebral disk calcifications were reported. SAAserum levels and other serological markers were measured too. Specimens were analysed by Congo Red, Immunofluorescence, Transmission Electron Microscopy.

Results: The analysis of all AKU specimens confirmed the massive presence of amyloid fibrils even in young patients. Alterations in collagen composition, strictly associated to amyloid bundles deposition, were observed especially in labial salivary gland, cartilage, tendons and infrapatellar fat pad. Histological analysis showed depletion of glycosaminoglycans in young patients, whereas, at light microscopy, calcification and fibrillation were observed only in elderly patients. Immunofluorescence assessed undoubtedly the presence of SAA in amyloid deposits in AKU, and we reported for the first time the finding of amyloid deposition in young AKU patients and in less common regions.

Conclusions: We provide the first detailed overview of amyloidosis in AKU. Overall, our findings depicted a novel biological framework underlining the pathological role of amyloidosis in several AKU tissues. Furthermore, we found that degradation of extra-cellular matrix AKU is not limited to elderly. The clinical burden of AKU may notably increase, since amyloidosis was found even in young AKU patients, whereas degeneration of cartilage and tendons was limited to older subjects.

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SAT0621

CLINICAL SIGNIFICANCE OF INTERLEUKIN-18 AND INTERLEUKIN -6 ON DISEASE COURSE OF ADULT-ONSET STILL'S DISEASE

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Background: Innate pro inflammatory cytokines interleukin (IL)–6 and IL-18 are critical for perpetuating the inflammatory processes in adult-onset Still's disease (AOSD),¹ and macrophage activation syndrome (MAS).²

Objectives: To assess the role of IL-6 and IL-18 in the pathogenesis of (AOSD) and MAS, and to investigate the clinical significance of serum cytokine profile with IL-6 and IL-18 in (AOSD) and MAS,

Methods: we analysed the serum IL-6 and IL-18 in patients with (AOSD) and compared them with the clinical features of (AOSD) 51 patients with (AOSD) including 17 patients with MAS were analysed. Levels of IL-6 and IL-18 were quantified in serum by enzyme linked immune sorbent assay. Results were compared with clinical features of (AOSD).

Results: Two distinct Adult Still's disease patient subsets based on their serum IL-6 and IL-18 levels were identified: an IL-18 dominant (IL-18/IL-6 1000) and an IL-6 dominant (IL-18/IL-6 1000). The IL-6 dominant subset had a significantly greater number of joints with active disease and higher serum levels of matrix metalloproteinase-3, whereas the IL-18-dominant subset was more likely to develop MAS. The cut off value of serum IL-18 to predict the development of MAS was 52000 pg/ml with 87.3% of sensitivity and 79.3% of specificity. The patients with IL-18 dominant pattern were likely to have monophasic or polycyclic disease course, whereas the patients with IL-6 dominant pattern were likely to have persistent disease course. Serum IL-18 levels in patients achieved remission decreased to the levels <1,000 pg/mL in inactive phase and normalised in remission phase. In contrast, serum IL-18 levels in patients experienced relapse during withdrawal of steroid within 12 months after disease onset demonstrated a sustained elevation of serum IL-18 levels >1,000 pg/mL during the inactive phase

Conclusions: Two subsets of patients with (AOSD), one which is prone for arthritis and another with prone for MAS, can be identified on the basis of their serum IL-6 and IL-18 levels. These two subsets appear to be characterised by certain distinct clinical features. Monitoring the cytokine profile with IL-18 and IL-6 might be useful to predict disease course. Furthermore, serum IL-18 levels reflect the biological activities of the immune system in (AOSD) and may predict the development of MAS and the prognosis of Adult Still's disease

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SAT0622

HIGH DOSE INTRAVENOUS METHYLPREDNISOLONE INDUCES RAPID IMPROVEMENT IN SEVERE UVEITIS: A MULTICENTER STUDY OF 129 PATIENTS

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Background: In uveitis rapid and effective remission-inducing therapy is mandatory to avoid irreversible damage.

Objectives: To evaluate the efficacy and safety of high-dose intravenous methylprednisolone (IVMP) pulse therapy in uveitis.

Methods: Multicentre study of 129 patients (203 eyes) with uveitis who received IVMP. The underlying diseases were: Idiopathic (n=30), Vogt Koyanagi Harada,²⁹ Behçet disease,¹⁹ Sarcoidosis,⁶ Multifocal Choroidopathy,⁴ Birdshot chorioretinopathy,² Acute posterior multifocal placoid pigment epitheliopathy,¹ Granulomatosis with polyangiitis,² Aortitis,¹ Immune thrombocytopenic purpura,¹ Rheumatoid arthritis,² Axial Spondylitis,⁶ Psoriatic arthritis,² Juvenile Idiopathic Arthritis,¹ Thrombophilia synd.,¹ Eales Disease,² Sympathetic Ophthalmia,² Multiple Sclerosis,² Relapsing Polychondritis,¹ Cogan's synd.,¹ Sjögren synd.,² Crohn's disease,¹ Reactive arthritis,¹ Toxic oil synd. and brain ischemia,¹ Raynaud's Disease and brain ischemia,¹ Pseudotumor,¹ Cataract surgery,¹ Herpes Simplex,¹ Varicella Zoster-associated acute retinal necrosis,¹ *T. pallidum*³ and *M.*