Abstract AB1086 – Figure 1

Note: the number above each bar represents the number of patients at that dose. The ACR Pediatric-30, -50, -70, and -90 responses were defined as an improvement of at least 30% (or 50%, 70%, 90% respectively) from baseline in at least 3 of the 6 signs and symptoms variables, with no more than 1 of the remaining variables worsening by >30%. JIA signs and symptoms variables: physician's global assessment of disease activity, CHAQ disability index score, CHAQ global assessment of well-being, number of joints with active arthritis, number of joints with limited range of motion, serum CRP or ESR.

Conclusions: Improvement in JIA signs and symptoms occurred at most assessments and by month 6, the percentage of patients with an ACR Pediatric-30, -50, -70, and -90 Response was 47.1%, 38.2%, 32.4%, and 17.6%. No new safety signals were identified for the well-characterised components of this fixed dosed JIA treatment, which was developed to reduce the risk of gastric ulcers.

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AB1087 PROLONGED RESPONSE WITH TUMOUR NECROSIS FACTOR ALFA INHIBITION IN A 5 YEAR OLD BOY WITH SEVERE MANIFESTATIONS OF IL-36 RECEPTOR ANTAGONIST DEFICIENCY (DITRA)

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Background: Deficiency of the interleukin (IL)–36 receptor antagonist (DITRA) is an autosomal recessive autoinflammatory syndrome caused by mutations in the IL36RN gene. Clinical manifestations of DITRA include recurrent episodes of generalised skin postulation, fever, systemic inflammation and leukocytosis. An uniformly effective treatment for DITRA has not yet been identified.

Objectives: We present a case of a 5 year old patient with DITRA with prolonged response with tumour necrosis factor alfa inhibition with adalimumab.

Methods: A five-year-old came to our dermatology clinic after worsening of a previous diagnosed plaque psoriasis, with an erythematous scaly dermatitis that extended throughout the trunk. Treatment with acitretin and cyclosporin were not effective and patient developed in few weeks a generalised erythroderma with

pustules covering almost every part of his body, including palms and soles. He was admitted for the onset of fever and irritability due to painful rubbing of the skin. Family history of recurrent fevers or psoriasis were not revealed. Parents were not consanguineous.

Complete blood count showed leukocytosis with neutrophilia and thrombocytosis, with an erythrocyte sedimentation rate (ESR) of 6 mm/hr and a C-reactive protein (CRP) of 8.4 mg/dl. Biochemistry panel revealed a mild elevation of liver enzymes without other abnormalities. Antinuclear antibody (ANA) and rheumatoid factor were negative with normal serum immunoglobulin and complement. Blood culture grew *E. Coli*, *S. Maltophilia* and *S. epidermidis*. Skin biopsy showed acanthosis and papillomatosis with perivascular polymorphous inflammatory cells. Genetic analyses showed a homozygous mutation in the IL36RN gene (pSer113Leu). No mutations were detected in IL1RN and CARD 14 genes.

Results: Treatment was initiated with intravenous methylprednisolone 2 mg/kg/day and subcutaneous anakinra 2 mg/kg/day. Cefotaxime and co-trimoxazole were added until blood cultures were negative. Although skins lesions improved during the following days and patient was finally discharged, symptoms reappeared when decreasing the steroid dose. Three months later adalimumab and methotrexate were started, allowing the patient to end treatment with corticoids without evidence of activity of the disease.

Abstract AB1087 – Figure 1

Conclusions: After incomplete response with anakinra, inhibition of tumour necrosis factor alfa resulted in a prolonged response in our patient with deficiency of the interleukin (IL)–36 receptor antagonist (DITRA).

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AB1088 CAPILLARY HEMOSIDERIN DEPOSITS OR EXTRAVASATIONS: A SUBTYPE OF HAEMORRHAGETHAT ACQUIRES SEPARATE ATTENTION IN QUANTITATIVE ANALYSIS OF NAILFOLD CAPILLAROSCOPY IN CHILDHOOD-ONSET SLE

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Background: Quality of images in nailfold capillaroscopy has improved in the last years by introduction of videocapillaroscopy. Microangiopathy, as observed in capillaroscopy of SLE-patients,^{1–3} can now be described by more detailed quantitative analysis. Recently, in a small cohort (n=22) of childhood-onset SLE (cSLE), we described capillary bleedings by two different subtypes: large haemorrhages and small point-shaped haemorrhages with a total count of resp. 0.2/1.5 per

analysed image per patient.⁴ These small point shaped haemorrhages have also been described as hemosiderin deposits.⁵

Objectives: To describe the reliability of assessment of two subtypes in capillary haemorrhages by inter-observer agreement.

Methods: Three observers, DS (paediatric rheumatologist with experience in capillaroscopy), AN (fellow paediatric rheumatology without experience in capillaroscopy) and MB (trainee in adult rheumatology with experience in capillaroscopy), scored capillaroscopy images from patients with Raynaud's phenomenon and with cSLE. The observers were blinded for patient name and diagnosis. The number of haemorrhages were scored per subtype. Hemosiderin deposits were defined as small point-shaped extravasations surrounding the capillary apex (see image). Large haemorrhages were defined according to the Atlas of Capillaroscopy.⁶ Reliability was calculated by the intra-class correlation coefficient (ICC) with 95% confidence interval (CI). Statistical analyses was performed by IBM SPSS Statistics version 24.

Results: Two-hundred images from 50 patients (diagnosed with Raynaud's phenomenon and/or cSLE) were scored by the three independent observers. ICC for the number of capillaries with hemosiderin deposits was 0.77 (95% CI 0.69–0.82). ICC for the number of large capillary haemorrhages was 0.97 (95% CI 0.96–0.98).

Abstract AB1088 – Figure 1

Conclusions: Reliability of the observation 'hemosiderin deposits' in nailfold videocapillaroscopy was good with an ICC of 0.77. This study shows that capillary haemorrhages can be described in 2 subtypes: 'large haemorrhages' and 'hemosiderin deposits' which are small point-shaped extravasations surrounding the capillary apex.

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AB1089

INVESTIGATION OF THE EFFICACY AND SAFETY OF SECUKINUMAB TREATMENT IN JUVENILE IDIOPATHIC ARTHRITIS SUBTYPES OF JUVENILE PSORIATIC AND ENTESITIS-RELATED ARTHRITIS: DESIGN OF A RANDOMISED, DOUBLE-BLIND, PLACEBO CONTROLLED, MULTICENTER STUDY

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Background: Secukinumab (AIN457), a fully human anti-interleukin-17A monoclonal antibody, has demonstrated a significant clinically meaningful efficacy on signs and symptoms, structure and function in adults with ankylosing spondylitis (AS)¹ and psoriatic arthritis (PsA)², both approved indications. These data support the proposed study in children with enthesitis-related arthritis (ERA) and juvenile psoriatic arthritis (JPsA).

Objectives: This phase 3 study will investigate the efficacy and safety of secukinumab in children ≥ 2 to < 18 years with active JPsA or ERA. The primary objective is to demonstrate that the time to flare in a double-blind placebo control treatment withdrawal part of the trial is longer with secukinumab than placebo.

Methods: Eighty biologic-naïve children with active ERA or JPsA (active ≥ 3 active joints and > 1 site of enthesitis at baseline or documented by history) will enrol into treatment period 1 and receive weekly open label s.c. secukinumab 75 or 150 mg, based on their body weight (< 50 kg or ≥ 50 kg) to maintain secukinumab blood levels equivalent to the adult 150 mg dose, for the first month then every 4 weeks thereafter. At week 12, responders (minimum JIA ACR Pedi 30 response) enter treatment period 2 and will be randomised to receive secukinumab or a matching placebo every 4 weeks. Patients enter treatment period 3 if they experience a disease flare or when the treatment period 2 closes for the entire study because the target number of flares has been reached. Upon entering treatment period 3, patients receive open-label secukinumab every 4 weeks until week 100 and then followed until week 112.

Results: The primary efficacy endpoint will be time to flare in treatment period 2. Key secondary endpoints include JIA ACR 30/50/70/90/100 response rate, total dactylitis and enthesitis counts at week 12. Safety and tolerability will be assessed throughout the study.

Conclusions: The efficacy of Secukinumab in the approved adult indications of PsA and AS support the current study design to evaluate the efficacy and safety of secukinumab treatment in children with active JPsA or ERA. The primary efficacy endpoint will be time to flare in treatment period 2. Key secondary endpoints include JIA ACR 30/50/70/90/100 response rate, total dactylitis and enthesitis counts at week 12. Safety and tolerability will be assessed throughout the study.

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AB1090

ANALYSIS OF A COHORT OF PATIENTS ATTENDING A COMBINED OPHTHALMOLOGY- RHEUMATOLOGY CLINIC IN A TERTIARY REFERRAL CENTRE IN EGYPT

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Background: Paediatric rheumatologic diseases may have sight-threatening ocular complications including uveitis, scleritis, and retinopathy. Conversely, children presenting with uveitis, scleritis, episcleritis, or optic neuritis may have an underlying rheumatologic disease. Combined ophthalmology-rheumatology clinics can facilitate the comprehensive management of these patients.

Objectives: To describe the demographic characteristics, paediatric rheumatologic diseases distribution, paediatric ocular manifestations distribution, and active treatments in a combined ophthalmology- rheumatology clinic in a tertiary referral centre in Egypt.