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.

REFERENCES:

- [1] Petty RE, et al. *J Rheumatol* 2004;31:390–392
- [2] Rigante D, et al. *Clinic Rev Allerg Immunol* 2015; 49:253–261
- [3] Ringold S, et al. *Arthritis Rheum* 2013; 65 (10):2499–2512

Disclosure of Interest: D. Lovell Grant/research support from: National Institutes of Health, NIAMS. Cincinnati Childrens Hospital Medical Centre receives funds from AstraZeneca, Bristol-Myers Squibb, AbbVie, Pfizer, Roche, Novartis, UBC, Forest Research Institute, Horizon Pharma, Johnson and Johnson, Biogen, Takeda, Genentech, GlaxoSmithKline, Boehringer Ingelheim, Celgene, and Janssen for consulting, Speakers bureau: Genentech and Bristol Meyers Squibb, J. Dare Grant/research support from: AbbVie, AstraZeneca, Bristol-Myers Squibb, Horizon Pharma, Medac, Pfizer, Roche and UCB, J. Ball Shareholder of: Horizon Pharma USA, Inc., Employee of: Horizon Pharma USA, Inc., M. Francis-Sedlak Shareholder of: Horizon Pharma USA, Inc., Employee of: Horizon Pharma USA, Inc., B. LaMoreaux Shareholder of: Horizon Pharma USA, Inc., Employee of: Horizon Pharma USA, Inc., R. Holt Shareholder of: Horizon Pharma USA, Inc., Employee of: Horizon Pharma USA, Inc.

DOI: 10.1136/annrheumdis-2018-eular.1089

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)

D. Clemente¹, J.C. López Robledillo¹, A. Torrolo², A. Hernández², E. Villalobos³.
¹Pediatric Rheumatology Unit; ²Pediatric Dermatology; ³Pediatrics, Hospital Niño Jesús, Madrid, Spain

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).

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6532

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

D. Schonenberg-Meinema¹, M. van den Berg¹, A. Nassar-Sheikh-Rashid¹, M. Boumans², M. Cutolo³, T. Kuijpers¹, V. Smith⁴, on behalf of EULAR study group on microcirculation in rheumatic diseases. ¹Pediatric Hematology, Immunology, Rheumatology and Infectious diseases, Emma Childrens Hospital, Academic Medical Center (AMC); ²Department of Clinical Immunology and Rheumatology, Academic Medical Center (AMC), Amsterdam, Netherlands; ³Research Laboratory and Academic Unit of Clinical Rheumatology, University of Genova, Genova, Italy; ⁴Department of Rheumatology, Ghent University Hospital, Ghent, Belgium

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