period (FUP, <12 months), while the corresponding response rates at long-term FUP (≥ 12 months) were 45%, 51%-63%, 58%-100% and 60%-100%. For ADA and ADA or ETN or INF (patients were on either therapy), while the JIA-ACR response rates at short term FUP were not reported, the response rates at long-term FUP were 100% and 78% respectively. Additionally, juvenile arthritis disease activity score (JADAS)-10 ≤ 1 scores were reported for ANA or CAN, CAN, ETN and TCZ as 52%, 48%-91%, 20% and 36% at short-term FUP. The mean changes in JADAS-71 scores at short-term FUP for ANA and TCZ were 10% and 11% respectively while the corresponding scores at long-term FUP were 13% and 14% ($p < 0.001$).

All interventions were generally well tolerated by SJIA patients; infections, injection site reactions and macrophage activation syndrome were reported for all biologics. Other complications included gastrointestinal disorders, pharyngitis, skin disorders, increase in liver enzymes etc.

Conclusion: The current interventions especially ANA, CAN, ETN and TCZ were found to be effective and generally well tolerated in SJIA. However, the lack of head-to-head studies limits a rigorous comparison.

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FRI0599

AA AMYLOIDOSIS IN RECIPIENTS OF CADAVERIC DERIVED PITUITARY GROWTH HORMONE – POTENTIAL EFFECT OF AN AMYLOID ENHANCING FACTOR?

Helen J. Lachmann¹, Dorota Rowczenio, Janet A. Gilbertson, Islam Noor, Hadija Trojer, Julian D. Gillmore, Ashutosh D. Wechalekar, Philip N. Hawkins. *UCL Division of Medicine and Royal Free Hospital London NHS Foundation Trust, National Amyloidosis Centre, London, United Kingdom*

Background: The development of Creutzfeldt-Jakob disease (CJD) in individuals treated with human cadaveric pituitary-derived growth hormone (c-hGH) was reported in 1985 resulting in its immediate withdrawal(1). Amyloid- β protein (A β) deposits have been found at autopsy in individuals previously exposed to c-hGH and misfolded A β peptide was recently identified in archived vials of human c-hGH; this induced A β pathology in an experimental model(2). The suggestion that contaminants in c-hGH could seed protein misfolding is reminiscent of the concept of amyloid enhancing factor (AEF), in which administration of tiny amounts of amyloidotic material is associated with acceleration of experimentally induced murine AA amyloidosis(3). In humans AA amyloid is a rare complication of chronic inflammation in which the amyloid fibrils are derived from Serum Amyloid A protein (SAA), a circulating component of the acute phase response.

Objectives: To seek evidence of prior exposure to c-hGH in patients with AA amyloidosis.

Methods: A database of 782 patients with AA amyloidosis seen between 1990 & 2017 was searched using: pituitary, craniopharyngioma & growth hormone. Only patients exposed to GH prior to 1985 were included.

Results: 10 cases were identified; accounting for 8% of AA of unknown etiology. 8 were female, 9 north European, median age at diagnosis of amyloid 47.9 yr with a median time since starting c-hGH of 35 yr. The indications were: 4 craniopharyngioma, resection of ependymoma, nasal pharyngeal tumour & encephalocele, 2 unexplained hypopituitarism. Chronic inflammation was assessed by serial blood tests after the diagnosis of amyloid; 1 had median SAA 131 mg/L; 4 had medians 20- 54

mg/L & the others low level inflammation, median < 20mg/L. No cause of inflammation was identified although 50% were obese. Genetic testing excluded systemic autoinflammatory disorders. 6 European patients were homozygous for SAA1.1 (66.7% compared to control frequency 29%)(4). 4 patients eventually required dialysis, 2 had renal transplants; median survival from diagnosis of amyloid was 132 mn & 3 patients have died.

Conclusion: 2,000 UK children were exposed to c-hGH; the finding of AA amyloid of uncertain etiology in these 10 cases raises the possibility that contaminants in c-hGH potentially contribute to misfolding of SAA. 80% had at least one recognised additional modest risk factor for the development of AA amyloidosis in the form of SAA1.1 homozygosity or obesity suggesting that a combination of factors may contribute to amyloidogenesis in individuals who have low levels of circulating potentially amyloidogenic protein(4).

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FIRST SINGLE-CENTERED COHORT OF CHINESE PATIENTS WITH PEDIATRIC SAPHO

Yuming Shao¹, Chen Li², Shuang Liu³, Wen Zhang⁴. ¹Peking Union Medical College, Beijing, China; ²Peking Union Medical College Hospital, Traditional Chinese Medicine, Beijing, China; ³Peking Union Medical College, Beijing, China; ⁴Peking Union Medical College Hospital, Rheumatology, Beijing, China

Background: The SAPHO syndrome (Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis) is an disease entity with chronic inflammatory osteo-articular symptoms and typical dermatological lesions. Pediatric SAPHO is regarded as equivalent of Chronic Recurrent Multifocal Osteomyelitis (CRMO) or Chronic Non-bacterial Osteomyelitis (CNO). There have already been several pediatric SAPHO cohorts about Caucasian populations. No Chinese cohorts have been reported.

Objectives: The aim of this study was to evaluate clinical features and treatment for all pediatric SAPHO patients admitted to Peking Union Medical College Hospital (PUMCH, Beijing, China)

Methods: We conducted a single-centered non-intervention retrospective study of 24 pediatric SAPHO patients who were diagnosed with Khan's modified criteria in PUMCH from April 2014 to August 2018 [1]. The demographic (sex, age, onset age, follow-up years), clinical (bone and skin symptoms), laboratory (ESR, CRP, HLA-B27, ANA, IL-6, IL-8, TNF- α), imaging (CT, MRI, bone scintigraphy), histologic (lymphocytic, granulocytic) and treatment (medications and effect) data were collected.

Results: Detailed information of 15 male and 9 female was available. The mean $\pm$ SD age at onset of bone and skin symptoms was 11.7 ± 3.8 and 14.4 ± 2.7 years, respectively. The duration of follow-up was 19.2 ± 15.2 months. 17 (71%) patients had skin manifestations (46% Severe Acne, all male; 21% Palmoplantar Pustulosis, all female; 4% psoriasis). Involvement of bone lesions varied (42% anterior chest wall, 29% mandible, 50% peripheral bones, 21% spinal bones). ESR, CRP, IL-6, IL-8, IL-10 was elevated in 88%, 96%, 50%, 71%, 70% patients, respectively. 11 did bone biopsy (6 dominantly lymphocytic, 3 granulocytic, 2 both). CT, MRI and bone scintigraphy were performed in 79%, 96%, 96% patients, respectively. A total of 6 patients have been treated with NSAIDs, 10 with bisphosphonate, 10 with TNF- α antagonist, 1 with Glucocorticoids, with a variable response. 70% patients showed complete remission after bisphosphonate or TNF- α antagonist therapy.