The most commonly (58.5%) used imaging method for diagnosis was computed tomography (CT). All the patients used initial glucocorticoid treatment. 4 patients (7.5%) received only glucocorticoid, others were underwent the following treatments combined with glucocorticoid: azathioprine (AZA) (60.4%); methotrexate (mtx) (11.3%), rituximab (RTX) + AZA (9.4%), mtx + AZA (5.7%), RTX (3.8%) and infliximab (1.9%).

Table 1. Distribution of clinical findings and tissue/organ involvement (n=53)

Constitutional symptoms	32 (22.4)
Rheumatoid arthritis	29 (34.7)
Cardiovascular disease**	
Pericarditis	24 (45.3)
Aortic	22 (39.6)
Aortic aneurysm	1 (1.9)
Pericardial effusion	9 (16.7)
Sclerodermatitis	6 (11.3)
Psoriasis	6 (11.3)
Sialadenitis	9 (16.7)
Sinusitis	9 (16.7)
Orbital pseudotumor	4 (7.5)
Lymphadenopathy	4 (7.5)
Arthritis	3 (5.7)
Fibrosing sialadenitis	3 (5.7)
Sclerosing cholangitis	3 (5.7)
Kidney disease	1 (1.9)
Tubulointerstitial nephritis	2 (3.8)
kidney mass	1 (1.9)
Thyroiditis	1 (1.9)
Encephalitis disease	1 (1.9)
Lung disease	1 (1.9)

**Two patients had pericarditis and aortic aneurysm and one patient had pericarditis and pericardial effusion together.

Table 2. Demographic and clinical features of patients

Parameter	Result
Age (year)*	51.49 ± 12.36
Gender (male) n (%)	27 (50.9)
Smoking (yes) n (%)	23 (43.4)
Comorbidity n (%)	
Hypertension	18 (34)
Diabetes mellitus	16 (30.2)
Chronic renal failure	7 (13.2)
Nephrotic syndrome	6 (11.3)
Autoimmune	6 (11.3)
Coronary artery disease	5 (9.4)
Chronic obstructive pulmonary disease	4 (7.5)
Malignancy	3 (5.7)
ESR (mm/h)*	50.7 ± 28.7
CRP (mg/L)*	35.5 ± 51.9
Cave (mg/dL)*	1.37 ± 1.52
CRP*	78.3 ± 32.7
IgG4 concentration (mg/dL)*	561.6 ± 907.6

*Continuous variables were presented as mean ± SD.

In the follow-up, a significant decrease in acute phase reactants was found in 62% of the patients at their last visits. While 27.3% of the patients had complete remission, 36.4% had partial remission, 20.5% had stable course, 13.6% had progression in the disease and 2.3% had recurrence. In 18 patients (64.3%) out of 28 patients who were in partial or complete remission, remission was achieved by using glucocorticoid and AZA combination treatment.

Conclusion: In conclusion, we have described a considerably large series of patients with IgG4-related disease from Turkey. The results of the study suggested that AZA and glucocorticoid combination treatment was commonly used in Turkish patients with IgG4-related disease and it might be a good treatment option to achieve remission.

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Disclosure of Interests: Aydan Köken Avcı: None declared, Önay Gerçek: None declared, Gerçek Can: None declared, Berrin Zengin: None declared, Sinem Burcu Kocær: None declared, Atilla Okan Kılıç: None declared, Sadettin Uslu: None declared, Ali Karakas: None declared, Merih Birlik: None declared, Servet Akar Grant/research support from: MSD, Abbvie, Roche, UCB, Novartis, Pfizer, Amgen, Consultant for: MSD, Abbvie, Roche, UCB, Novartis, Pfizer, Amgen, Speakers bureau: Pfizer, Fatos Onen: None declared

DOI: 10.1136/annrheumdis-2019-eular.4632

FRI0597

INVOLVEMENT OF TWO OR MORE SETS OF LACRIMAL GLANDS AND/OR MAJOR SALIVARY GLANDS IS RELATED TO GREATER SYSTEMIC DISEASE ACTIVITY IN IGG4-RELATED DACRYOADENITIS/SIALADENITIS

Masahiro Konishi, Ichiro Mizushima, Hajime Sanada, Kazuyuki Suzuki, Akari Takeji, Satoshi Hara, Kiyoaki Ito, Hiroshi Fujii, Kazunori Yamada, Mitsuhiro Kawano. Kanazawa University Hospital, Department of Rheumatology, Kanazawa, Japan

Background: IgG4-related dacryoadenitis/sialadenitis (IgG4-DS) is a major organ manifestation of IgG4-related disease (IgG4-RD) and is notable for its lack of a marked male predominance [1]. In IgG4-DS, involvement of two or more sets of lacrimal glands (LGs) and/or major salivary glands (MSGs) is regarded as a specific finding with diagnostic significance [2]. However, the influence of the presence of this finding on clinical features of IgG4-DS has not been well evaluated.

Objectives: This study aimed to clarify the influence of the involvement of ≥ 2 sets of LGs and/or MSGs on clinical features of IgG4-DS.

Methods: We retrospectively reviewed the medical records of 130 patients with IgG4-RD at a single center in Japan. Among them, 97 patients were diagnosed with IgG4-DS. We divided the 97 patients into Group A (n=44) with involvement of ≥ 2 sets of LGs and/or MSGs and Group B (n=53) with involvement of one set of LGs or MSGs or unilateral gland involvement. We compared the clinical features of Group A and B. Moreover, 33

IgG4-DS patients without extra-lacrimal/salivary gland lesions (DS-limited patients) were divided into Group a (n=14) and b (n=19) according to the presence/absence of the involvement of ≥ 2 sets of LGs and/or MSGs respectively, and their clinical features were also compared.

Results: The IgG4-DS patients comprised 60 men and 37 women (mean age 64.1 years). The mean follow-up period was 61.6 months, and the mean serum IgG4 level at diagnosis was 757 mg/dL. Group A had significantly more affected organs (3.5 ± 1.6 vs 2.7 ± 1.6 , $P=0.010$), lower serum C3 and C4 levels (83.6 ± 29.5 vs 95.0 ± 25.2 , $P=0.018$, and 16.5 ± 9.8 vs 21.4 ± 9.5 , $P=0.034$, respectively), and a tendency to have higher serum IgG levels ($2,509 \pm 1,066$ vs $2,165 \pm 911$, $P=0.075$) and IgG4-RD responder index (14.7 ± 6.7 vs 12.2 ± 6.5 , $P=0.050$) than did Group B. In the 33 DS-limited patients, compared with Group b, Group a had no significant differences in clinical features including the proportion of patients with serum IgG4 elevation [$\geq$ upper limit of normal (ULN), 85.7% vs 78.9%, $P=1.000$; $\geq 2 \times$ ULN, 64.3% vs 47.4%, $P=0.482$; $\geq 5 \times$ ULN, 28.6% vs 21.1%, $P=0.695$].

Conclusion: Involvement of two or more sets of lacrimal glands and/or major salivary glands suggests greater systemic disease activity mainly due to involvement of more organs.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5628

FRI0598

A SYSTEMATIC REVIEW OF THE EFFECTIVENESS AND SAFETY OF PHARMACOLOGICAL TREATMENTS IN PATIENTS WITH SYSTEMIC IDIOPATHIC JUVENILE ARTHRITIS (SJIA)

Jasmin Kuemmerle-Deschner¹, Lalit Thakur², Aneesh George², Peter Hur³.

¹Division of Pediatric Rheumatology, Department of Pediatrics, University Hospital, Tuebingen, Germany; ²Novartis Healthcare Pvt. Ltd., Hyderabad, India; ³Novartis Pharmaceuticals Corporation, East Hanover, New Jersey, United States of America

Background: Treatment options for SJIA include canakinumab, tocilizumab (both approved by US FDA and EMA) and anakinra (approved by EMA), while anti-tumor necrosis factor inhibitors, steroids, disease-modifying anti-rheumatic drugs and non-steroidal anti-inflammatory drugs are also used.¹ A recent systematic review included only randomized controlled trials (RCTs) of biologics. We aimed to generate a more comprehensive evidence on the effectiveness and safety of treatments used in SJIA.

Objectives: To assess evidence on the effectiveness and safety of therapeutic agents for SJIA from RCTs and real-world studies.

Methods: A systematic literature review was conducted using Cochrane methodology² from 2000 to Jan 2018. Sources included Embase®, MEDLINE®, MEDLINE®-In Process and Cochrane library. Studies were searched for English language publications as full-text articles (2000 to Jan 2018) or conference abstracts (2015 to Jan 2018). Studies with ≤ 15 pts were excluded.

Results: Of the 62 included studies, 8 were RCTs and 54 were real-world studies. The interventions included in the RCTs were anakinra (ANA), canakinumab (CAN), etanercept (ETN), methotrexate (MTX) and tocilizumab (TCZ) (1 RCT each and 1 withdrawal trial for TCZ), and 2 RCTs for rilonacept (RLN); all vs. placebo (PLB). In addition to the interventions mentioned in the RCTs, real-world studies also included other interventions such as abatacept, adalimumab (ADA), infliximab (INF), non-steroidal anti-inflammatory drugs and steroids. Juvenile idiopathic arthritis - American college of rheumatology (JIA ACR)-30 was the most common composite outcome reported across studies.

In RCTs, the JIA ACR-30 (+ no fever) responses were significantly superior vs. PLB for CAN (81% vs. 10%), TCZ (85% vs. 24%), and ANA (92% vs. 50%) ($p < 0.05$ for all), while other interventions (RLN and MTX) were less responsive. Additionally during the trial extension period, the inactive disease rates were 57%, 31% and 31% for TCZ, CAN and ANA respectively.

In real-world studies, the JIA-ACR 30 response rates reported for ANA, ANA or CAN (patients were on either therapy), TCZ and ETN were 50%-55%, 57%-60%, 47%-100% and 47%-73% at short-term follow-up

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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Disclosure of Interests: Jasmin Kuemmerle-Deschner Grant/research support from: Jasmin Kuemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis., Consultant for: Jasmin Kuemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis., Speakers bureau: Jasmin Kuemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis., Lalit Thakur Employee of: Lalit Thakur is an employee of Novartis Healthcare Pvt. Ltd., Hyderabad, India, Aneesh George Employee of: Aneesh George is an employee of Novartis Healthcare Pvt Ltd., Peter Hur Employee of: Peter Hur is an employee of Novartis Pharmaceuticals Corporation, East Hanover, New Jersey, USA

DOI: 10.1136/annrheumdis-2019-eular.5668

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 amyloidogenic 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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Disclosure of Interests: Helen J. Lachmann Grant/research support from: SOBI, Novartis, Consultant for: Novartis, Takeda, Speakers bureau: SOBI, Novartis, Dorota Rowczenio: None declared, Janet A. Gilbertson: None declared, Islam Noor: None declared, Hadija Trojer: None declared, Julian D. Gillmore: None declared, Ashutosh D. Wechalekar: None declared, Philip N. Hawkins: None declared

DOI: 10.1136/annrheumdis-2019-eular.2653

FRI0600

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 $\pm$ 3.8 and 14.4 $\pm$ 2.7 years, respectively. The duration of follow-up was 19.2 $\pm$ 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.