Objectives: To identify predictors of relapse of IgG4-RD after induction therapy.

Methods: We retrospectively reviewed 57 patients diagnosed with IgG4-RD and treated with GC in our hospital between January 2004 and November 2018. Clinical features at baseline, including organ involvement and blood markers (total hemolytic complement [CH50], its fractions [C3 and C4], IgG4, IgG, IgE, anti-nuclear antibody, rheumatoid factor, C-reactive protein, soluble interleukin-2 receptor, eosinophils), were collected. We divided patients into 2 groups on the basis of clinical features and examined whether they relapsed. In this study, hypocomplementemia was defined as decreased serum C3, C4, or CH50 less than the lower limit of normal. A relapse was defined as any new or worsened state of disease activity that required an escalation in treatment (immunosuppressants and/or GC). The follow-up period was defined as 182 days.

Results: Forty-three men and 14 women (mean age 68.1 ± 10.9 years) were included. Both serum IgG4 and IgG were measured at baseline in all patients (mean IgG4 798.8 ± 873.1 mg/dL and mean IgG 2874.0 ± 1934.1 mg/dL, respectively). All of the serum C3, C4, and CH50 were measured at baseline in 34/57 patients (mean C3 75.7 ± 33.3 mg/dL; mean C4 14.8 ± 11.5 mg/dL; and mean CH50 37.0 ± 23.1 U/mL). Fifteen patients had at least one episode of hypocomplementemia, and 19 patients did not. Most patients had multiple organ lesions. The details of dominant lesions were as follows: Dacryoadenitis and/or sialadenitis (Mikulicz disease), 36/57 patients (63.2%); biliary or pancreatic lesion, 29/57 (50.1%); retroperitoneal fibrosis, 22/57 (38.6%); and renal lesion 16/57 (28.1%). All patients were given prednisolone and gradually reduced (mean induction dose 31.7 ± 9.8 mg/day). No patients received immunosuppressant as induction therapy. During the follow-up period, 6 patients relapsed, and the dose of prednisolone was increased immediately. Relapsed lesions were as follows: Mikulicz disease, 3/6 patients (50.0%); biliary or pancreatic lesion, 1/6 (16.7%); retroperitoneal fibrosis 1/6 (16.7%); and pulmonary lesion 1/6 (16.7%). Patients with hypocomplementemia had significantly shorter relapse-free survival than those without ($p=0.039$, Figure 1). Patients with decreased serum C4 or CH50 (less than the lower limit of normal) also had significantly shorter relapse-free survival than those without, but those with decreased serum C3 did not. Such a tendency was not seen in other blood markers (IgG4, IgG, IgE, anti-nuclear antibody, rheumatoid factor, C-reactive protein, soluble interleukin-2 receptor, eosinophils).

Conclusion: Hypocomplementemia at baseline in patients with decreased serum C4 or CH50 may predict relapse of IgG4-RD after prednisolone therapy.

REFERENCES:

- [1] Stone JH, et al. N Engl J Med 2012; 366: 539-51.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2391

THU0593

CLINICAL AND GENETIC FEATURES OF CHINESE ADULT PATIENTS WITH TUMOR NECROSIS FACTOR RECEPTOR-ASSOCIATED PERIODIC SYNDROME

Mengzhu Zhao, Yi Luo, Min Shen, Di Wu, Wen Zhang, Xiaofeng Zeng. Department of Rheumatology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Key Laboratory of Rheumatology and Clinical Immunology, Ministry of Education, Beijing, China

Background: Tumor Necrosis Factor Receptor-Associated Periodic Syndrome (TRAPS) is an autosomal dominant autoinflammatory disease, associated with the mutation of tumor necrosis receptor superfamily member 1A (*TNFRSF1A*) gene,

located on chromosome12p13. TRAPS is usually diagnosed during pediatric age. However, adult-onset disease or diagnosis during adulthood has been occasionally described. Moreover, TRAPS has been hardly reported in the Chinese population. Herein, we aimed to characterize the clinical and genetic features of Chinese adult patients with TRAPS.

Objectives: Adult patients (≥ 16 years) suspected monogenic autoinflammatory diseases during the period April 2015 to October 2018, at the adult autoinflammatory disease center, Department of Rheumatology, Peking Union Medical College Hospital (PUMCH).

Methods: Clinical data were evaluated. Gene sequencing was performed in each patient to support the diagnosis and exclude other monogenic autoinflammatory diseases. Finally we compared the data with those from Japan and Europe.

Results: During the study period, 8 patients with TRAPS were diagnosed and follow-up. The ratio of male to female was 3: 1. The median age of disease onset was 4 (0.5–38.5), and adult-onset was observed in 2 (25%) patients. The median time of diagnosis delay was 17.8 (1.5–50.5) years. There were seven Chinese Han and one Manchu patients. One patient had a family history of TRAPS. The most frequent symptom was fever (8, 100%). The attacks of 5 (62.5%) patients lasted more than 1 week, and the intervals of 7 (87.5%) patients were longer than 2 weeks. Skin involvement occurred in 6 (75%) patients, with maculopapular mentioned in 6 and erythema annulare presented in 3. Four (50%) patients had arthralgia, while one had polyarthritis. 4 (50%) patients complained respectively of myalgia or headache. 4 (50%) patients experienced abdominal pain and one (12.5%) had vomiting. One (12.5%) patient had periorbital edema, while 3 (37.5%) had conjunctivitis. Up to 5 (62.5%) patients got nonspecific pharyngitis. No patient suffered from chest pain or amyloidosis. Eight gene variants were detected in *TNFRSF1A* gene. Heterozygous gene variants were found in 7 Chinese Han patients, and homozygous (c.769-23, T>C) happened to the Manchu patient. The variants included C58 (exon 2), G65E (exon 3), F89L (exon 3), C99G (exon 3), V202G (exon 6), c.769-23T>C (IVS8), S290I (exon 9) and m.1735A>G (non-coding region). NSAIDs were given to 2 patients, glucocorticoid or immunosuppressive agents given to 3 patients respectively, and etanercept to 5 patients. A complete response was found in all the 5 patients received etanercept. The effectiveness of other drugs were 50% in NSAIDs, 66.7% in glucocorticoid and immunosuppressive agents.

Conclusion: This is the first and largest case series of TRAPS in Chinese adult patients. It highlights the importance of screening *TNFRSF1A* gene in patients with unexplained periodic fever syndrome. Four novel *TNFRSF1A* variants, S290I, F89L, V202G and m.1735A G in non-coding region, have been identified. The atypical clinical manifestations of our patients compared to those from Europe might be related to their low-penetrance *TNFRSF1A* variants. Further studies are needed to explore the more accurate phenotypes and genotypes of the Chinese patients with TRAPS.

Figure legends: clinical manifestations of Chinese adult patients with TRAPS. A. Periorbital edema; B. erythema annulare; C. Maculopapular rash; D. Radiology showed intestinal obstruction.

Table 1 Summary of the main clinical and genetic features of the 8 Chinese adult patients

with TRAPS								
Patient	1	2	3	4	5	6	7	8
Sexuality	M	F	M	F	M	M	M	M
Nationality	Han	Han	Han	Han	Han	Han	Manchu	Han
Age at onset (years)	0.5	17	5	5	2.5	2	38.5	3
Age at diagnosis (years)	17	24	24	25	16	52	40	27
Diagnosis delay (years)	16.5	7	19	20	13.5	50	1.5	24
Family History	-	-	-	+	-	-	-	-
TNFRSF1A	m.1735A	G65	S29	C58	F89L	V202	c.769-23[IVS8]T>C	C99
Variant	>G	E	OI	R	-	G	-	G
Variant in other genes	-	-	-	-	R202Q in MEFV	-	D424E in MEFV, G39V in NLRP12	-
Fever	+	+	+	+	+	+	+	+
Duration	>3w	1w	2-3 d	1-2 w	1-2w	2-3d	2-3w	1w
Interval	<1w	3m	1-2 m	≥1 m	≥1m	<1m	>1w	1m
Rash	+	-	-	+	+	+	+	+
Periorbital edema	-	-	-	-	-	-	-	+
Conjunctivitis	-	-	+	-	-	+	+	-
Arthralgia/Arthritis	+	-	+	-	-	+	-	+
Myalgia	+	-	-	+	+	+	+	-
Abdominal pain	-	+	-	+	-	+	-	+
Angina	-	+	-	+	-	+	+	+
Headache	+	-	-	-	-	+	+	+

Table 2 Comparison of phenotypic and genotypic features among reported cohorts

Cohorts	PUMCH (n=8)	Japan (n=51)	Europe (n=158)
M : F	6:2	22:29	78:80
Young onset (<18 years old, %)	87.5	68.1*	78
Age at onset (years)	4 (0.5-38.5)	ND*	4.3 (0 - 63)
Age at diagnosis (years)	24.5 (14 - 52)	ND	25.9 (0-77)
Diagnosis delay (years)	17.8 (1.5 - 50.5)	ND	10.3 (0-77)
Family history (%)	12.5	45.1	ND
Fever (>38°C, %)	100	100	84
Rash (%)	75	55	63
Periorbital edema (%)	12.5	9.1	20
Conjunctivitis (%)	37.5	18.2	22
Arthralgia/Arthritis (%)	50	59	64
Myalgia (%)	54	43	70
Abdominal Pain (%)	50	36	70
Pharyngitis (%)	62.5	ND	21
Headache (%)	50	23	23
Diarrhea (%)	12.5	ND	18
Chest pain (%)	0	13.6	25
Amyloidosis (%)	0	0	10

*Age onset < 20 years old; *ND: No Data

REFERENCES:

- [1] Lachmann HJ, et al. The phenotype of TNF receptor-associated autoinflammatory syndrome (TRAPS) at presentation: a series of 158 cases from the Eurofever/EUROTRAPS international registry. *Ann Rheum Dis* 2014, 73: 2160-2167.
- [2] Ueda N, et al. Clinical and Genetic Features of Patients With TNFRSF1A Variants in Japan: Findings of a Nationwide Survey. *Arthritis Rheumatol* 2016, 68: 2760-2771.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5204

Diagnostics and imaging procedures

THU0594

CLINICAL VERSUS IMAGING REMISSION IN JUVENILE IDIOPATHIC ARTHRITIS (JIA): PRELIMINARY RESULTS OF THE REMECO STUDY

Marta Mazzoni^{1,2}, Silvia Merlo¹, Angela Pistorio³, Stefania Viola², Alessandro Consolaro^{1,2}, Angelo Ravelli^{1,2}, Clara Malattia^{1,2}. ¹Università degli studi di Genova, Genova, Italy; ²IRCCS Istituto Giannina Gaslini, Clinica Pediatrica e Reumatologia, Genova, Italy; ³IRCCS Istituto Giannina Gaslini, Epidemiologia e Biostatistica, Genova, Italy

Background: remission is becoming a realistic target in JIA, but clinical remission (CR) may not accurately reflect real absence of synovitis. It would be desirable to have instruments to predict the risk of relapse in patients in CR in order to establish the most appropriate therapeutic strategy. Despite in RA the role of imaging to predict disease flare is established, this field has remained almost unexplored in JIA.

Objectives: 1) to investigate the prevalence of musculoskeletal ultrasound (MSUS)-detected subclinical synovitis in JIA patients in CR; 2) to establish which and how many joints should be scanned to reliably assess remission; 3) to evaluate the persistence of subclinical synovitis over the time; 4) to investigate whether subclinical synovitis entails a risk of disease flare; 5) MSUS data will be integrated with serum levels of inflammatory biomarkers to develop a multidimensional measure of remission.

Methods: it is a longitudinal prospective 4 years study started on November 2017. So far we have enrolled 99 consecutive JIA patients who met the Wallace criteria for CR. For each patient 46 joints were scanned for synovial hyperplasia/joint effusion and PD signal, all graded semiquantitatively on a 0-3 scale independently by 2 expert ultrasonographers. Subclinical synovitis was defined when total synovitis score for each joint was ≥2. MSUS was performed at baseline and at 6 month follow up visit. At inclusion serum assays have been stored to determine levels of inflammatory biomarkers (S100A8/9-A12, bFGF, IL-6, IL-10, CXCL9-10, VEGF, YKL40). A flare of synovitis was defined as a recurrence of clinically active arthritis.

Results: 99 patients (79.8% F; median age 11.3 y; median disease duration 5.3 y; median CR duration 1.6 y) were included. Thirty-eight/99 (38.4%) patients had persistent oligoarthritis; 34/99 (34.3%) extended oligoarthritis; 22/99 (22.2%) polyarthritis; 5/99 (5.1%) systemic arthritis. Fifty-nine/99 (59.6%) patients were in CR on medication. Subclinical synovitis was detected in 54/99 (54.5%; 95% CI: 45.2 - 65.5%) patients, PD in 18/99 (18.2%; 95% CI: 11.1 - 27.2%) patients; subclinical tenosynovitis in 7/99 (7.1%; 95% CI: 2.9 - 14%) patients. Subclinical synovitis was found more frequently in the ankle [31/54 (57.4%) patients] and wrist joints [17/54 (31.5%) patients]. No patients had subclinical synovitis in the hip. A 14-joint reduced count including bilateral knee, ankle (tibiotalar, subtalar and talonavicular joints), wrist (radiocarpal and intercarpal joints) and elbow joints, detected 92.6% of children with subclinical synovitis. Twenty-five/99 (25.2%) patients in persistent CR were reassessed with MSUS at a follow up visit (median follow up duration 7 months): 82.3% of patients showed persistent subclinical synovitis. Sixty-four/99 (64.6%) patients had a clinically follow up of at least 6 months and 9/64 (14%) patients experienced a disease flare (median time to flare 6.6 months). Six/9 (66.7%) patients who experienced a relapse had subclinical synovitis at baseline.

Conclusion: our preliminary results confirm the discrepancy between clinical and imaging remission and that clinical evaluation may not sensitive to detect an inflammation-free state. Bilateral US assessment of the elbow, wrist, knee and ankle joints is reliable to detect subclinical synovitis. So far, patients who have relapsed are a small percentage, but to extend follow up is crucial to test predictive value of MSUS. Imaging findings will be combined with serum biomarkers leading to the construction of a predictive model.

REFERENCES:

- [1] De Lucia O, et al. Baseline ultrasound examination as possible predictor of relapse in patients affected by juvenile idiopathic arthritis (JIA). *Ann Rheum Dis*. 2018 Oct;77(10):1426-1431.

Disclosure of Interests: Marta Mazzoni: None declared, Silvia Merlo: None declared, Angela Pistorio: None declared, Stefania Viola: None

declared, Alessandro Consolaro Grant/research support from: AbbVie, Pfizer, Angelo Ravelli Grant/research support from: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Consultant for: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Speakers bureau: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Clara Malattia: None declared

DOI: 10.1136/annrheumdis-2019-eular.7082

THU0595 DEVELOPMENT OF AN AUTOMATED SEGMENTATION ALGORITHM TO IDENTIFY BONES OF THE HAND

Adwaye Rambojun¹, William Tillett^{2,3}, Neill Campbell⁴, Tony Shardlow¹. ¹University of Bath, Mathematical Sciences, Bath, United Kingdom; ²University of Bath, Pharmacology, Bath, United Kingdom; ³Royal National Hospital For Rheumatic Diseases, Bath, United Kingdom; ⁴University of Bath, Computer Science, Bath, United Kingdom

Background: The evaluation of structural damage with plain radiography is important to clinicians and patients. Standard scoring methods include the Sharp-van der Heijde (SVdH) and Raitingen methods [1] however these systems are time-consuming. Therefore, it is difficult to perform large cohort studies. We set out to develop an automated algorithm to identify bones on plain radiographs as a step towards developing automated quantification of structural damage for use on large datasets.

Objectives: To develop a novel algorithm to segment outlines of finger bones in hand radiographs.

Methods: 101 hand radiographs were gathered from the Bath longitudinal cohort (UK). All patients fulfilled the CASPAR criteria for Psoriatic Arthritis (PSA). None of the patients had damage on SVdH and Raitingen scoring (blinded). The metacarpal (MC), proximal phalanx (PP), middle phalanx (MP), and distal phalanx (DP) in the right index finger were delineated by a rheumatologist. These outlines were used to build a statistical model of the shape using a Gaussian Process Latent Variable Model (GPLVM) [2]. Bones are segmented by matching the shape on a radiograph to the statistical model.

Results: The performance of the matching algorithm was compared with a traditional algorithm (snakes) using the Adjusted Rand Score (ARND). The ARND score measures the similarity of the segmentation with the ground truth. A perfect segmentation has a score close to 1. We tested the algorithm on 9 PP, 9 MP and 8 DP and 6 MC bones in the right index finger. The results are reported in table 1. We report a mean improvement in ARAND of 0.19, 0.87, 0.43 and 0.30 for the PP, MP, DP and MC respectively.

Conclusion: We report a reliable algorithm for the identification of metacarpal, proximal, middle and distal phalanx bones of the hand. Future work will focus on using the output of the segmentation algorithm to track damage progression over time.

REFERENCES:

- [1] Van der Heijde, D., PAULUS, H., and SHEKELLE, P. (2000). How to read radiographs according to the Sharp/vander Heijde method. Discussion: Heterogeneity in rheumatoid arthritis radiographic trials. Issues to consider in a metaanalysis. *Journal of rheumatology*, 27(1):261–263.
- [2] Titsias, M. K. and Lawrence, N. D. (2010). Bayesian Gaussian process latent variable model. In *International Conference on Artificial Intelligence and Statistics*, pages 844–851.

Table 1. Adjusted RAND scores for comparing our algorithm to a traditional one (snakes)

	Bone	Snakes ARAND	Shape matching ARAND
Case 1	PP	0.70	0.95
Case 2	PP	0.89	0.96
Case 3	PP	0.82	0.96
Case 4	PP	0.72	0.94
Case 5	PP	0.53	0.96
Case 6	PP	0.87	0.97
Case 7	PP	0.79	0.96
Case 8	PP	0.74	0.96
Case 9	PP	0.88	0.97
Case 1	MP	0.79	0.95
Case 2	MP	0.75	0.95
Case 3	MP	0.75	0.94

Case 4	MP	0.72	0.95
Case 5	MP	0.77	0.96
Case 6	MP	0.51	0.96
Case 7	MP	0.76	0.96
Case 8	MP	0.77	0.96
Case 9	MP	0.83	0.97
Case 10	MP	0.77	0.93

Key: Adjusted Rand Score (ARND) score measures the similarity of the segmentation with the ground truth. A perfect segmentation has a score close to 1. Metacarpal (MC), proximal phalanx (PP), middle phalanx (MP), and distal phalanx (DP)

Figure 1. Shape matching algorithm output demonstrating segmented outlines of the DP, MP, PP and MC in red, green, orange, and blue respectively.

Disclosure of Interests: Adwaye Rambojun: None declared, William Tillett Grant/research support from: AbbVie, Celgene, and Lilly, Consultant for: AbbVie, Celgene, Lilly, Novartis, and Pfizer, Speakers bureau: AbbVie, Celgene, Lilly, Janssen, Novartis, UCB, and Pfizer, Neill Campbell: None declared, Tony Shardlow: None declared

DOI: 10.1136/annrheumdis-2019-eular.2336

THU0596 DIAGNOSTIC VALUE OF ULTRASOUND AND DUAL ENERGY COMPUTED TOMOGRAPHY TO ACHIEVE ACR-EULAR GOUT CLASSIFICATION CRITERIA IN REAL LIFE CLINICAL PRACTICE

André Ramon¹, Marie Schmitt¹, Romaric Ne², Pierre Emmanuel Berthod², Hervé Devilliers³, Jean Francis Maillefer¹, Paul Ornetti¹. ¹Dijon University Hospital, Rheumatology, Dijon, France; ²Dijon University Hospital, Radiology, Dijon, France; ³Dijon University Hospital, Internal Medicine, Dijon, France

Background: 2015 ACR/EULAR gout classification criteria (1) include ultrasound with double contour (DC) sign as key ultrasound features and dual energy computed tomography (DECT) with evidence of urate deposition. The positivity of either DECT or ultrasound allows 4 points in addition to others clinical and biological criteria to classify as gout is $\geq 8/23$. However, in routine care, the imaging modality that should be promoted remains unclear between ultrasound or DECT.

Objectives: To validate a possible diagnostic algorithm for the clinical use of DECT and ultrasound in suspected gouty arthritis.

Methods: We conducted a single-center prospective study in the Rheumatology Department of Dijon University Hospital from July 2016 to December 2018, including all patients hospitalized for suspected gouty arthritis. Each patient received joint aspiration if possible, an ultrasound assessment (DC sign and/or tophus) and DECT scanning of symptomatic joints. All these examinations were performed blind of the clinical data and results of joint aspiration. The gold standard used for this study was the 2015 ACR/EULAR gout classification criteria. We have established two