After the shooting, a website was created where the pictures were uploaded with underlined personal remarks of both the artists and the professional photographer. Information about the inflammatory rheumatic diseases and the importance of early diagnosis was added to the album. Additionally, during October 2017, a 45 s video was promoted through TV, targeting the serious problems that people with RMDs are facing in their everyday life but also revealing the strength and courage of this great artists' team. A press conference was as well organised to support the visit of the particular website.

Results: Results: 173.000 people have visited the website from October to January, with another 16.800 visits through YouTube.

Conclusions: Through an artistic approach a patient organisation can trigger the awareness of the public, communicate the important message of the seriousness of RMDs and also underline the endless dedication and the seek for dreams to come true, even if one has to live along with a rheumatic disease all your life.

Disclosure of Interest: None declared

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WEDNESDAY, 13 JUNE 2018:

Let's improve diagnosis and treatment of orphan diseases

OP0081 PHENOTYPIC SUBGROUPS IN IGG4-RELATED DISEASE – A CLUSTER ANALYSIS

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Background: IgG4-related disease (IgG4-RD) is a multi-organ immune-mediated condition of uncertain etiology characterised by substantial organ-specific morbidity if not diagnosed and treated promptly. Identifying IgG4-RD subgroups based on the distribution of organ involvement may influence the understanding of pathogenesis and guide clinical management.

Objectives: To identify phenotypic clusters of IgG4-RD that may differentiate clinically meaningful subgroups using an unbiased method.

Methods: The study cohort consisted of 493 IgG4-RD subjects diagnosed by 76 IgG4-RD specialists from North America, South America, Europe, and Asia. For each case, investigators included details regarding age at disease onset and diagnosis, race/ethnicity, organ involvement, biopsy findings, and lab results. We performed latent class analysis (LCA) using SAS procedure PROC LCA to identify subgroups representing distinct patterns of organ involvement by IgG4-RD (figure 1). We fitted LCA models with 2–5 subgroups and chose the best model based on Akaike information criteria and adjusted Bayesian information criterion. The posterior probability of subgroup (cluster) membership for all cases was determined and cases were assigned to the cluster in which they had the highest probability of membership. We compared the distribution of organ involvement and other baseline features between clusters using Chi square tests and analysis of variance, where appropriate.

Results: Of the 493 IgG4-RD subjects, 65% were male, 40% were Caucasian, 45% were Asian, and 12% were Hispanic. The mean age at diagnosis was 59.5 (±14.0) years. Using LCA, we identified four clusters of IgG4-RD (table 1), each of which accounted for between 19% and 32% of the cohort. Cluster 1

('Hepatobiliary') included 158 (32%) patients characterised by hepatobiliary involvement. Cluster 2 ('Orbital') included 88 (19%) patients characterised by orbital and/or sinus disease. Cluster 3 ('Mikulicz') included 109 (22%) patients who had features of classic Mikulicz (dacryoadenitis plus major salivary gland involvement), often accompanied by renal and lung disease. Cluster 4 ('Retroperitoneal Fibrosis (RPF)') included 138 (28%) patients with RPF and/or aortic involvement. The clusters differed significantly with regard to age at symptom onset ($p<0.001$), gender and race distribution ($p<0.001$), serum IgG4 concentration ($p=0.02$), and presence of hypocomplementemia ($p<0.001$). In contrast to the other clusters, cluster 2 ('Orbital') included a majority of female patients who tended to be younger. Cluster 3 ('Mikulicz') was characterised by the highest serum IgG4 concentrations and cluster 4 ('RPF') by the lowest. Hypocomplementemia, which occurred in only a minority of patients overall (9%), tended to segregate in cluster 3 ('Mikulicz'), a group in which renal disease was common.

Abstract OP0081 – Table 1

Variable (mean ±SD or median [IQR])	Cluster 1	Cluster 2	Cluster 3	Cluster 4	P-Value
Sample Size	158 (32%)	88 (19%)	109 (22%)	138 (28%)	
Organ Involvement	Hepatobiliary	Orbital - Sinusitis	Mikulicz - Renal - Lung	RPF - Aorta	
Male	79%	35%	62%	72%	<0.0001
Age at Diagnosis	62 (12.8)	56 (14.1)	60.1 (12.9)	58.3 (14.5)	0.008
Age at Symptom Onset	61.2 (13.0)	53.3 (14.5)	57.7 (13.9)	56.5 (15.7)	0.0004
Diagnostic Delay	0.87 (1.7)	2.7 (3.7)	2.5 (3.8)	1.9 (3.9)	<0.001
Race/Ethnicity					
White	46%	35%	19%	53%	<0.0001
Asian	39%	52%	51%	31%	
Hispanic	10%	9%	20%	9%	
Number of Organs	2.2 (1.0)	2.3 (1.3)	5.0 (1.9)	2.3 (1.3)	<0.0001
IgG4 (mg/dl) Concentration	325 (160, 667)	334.5 (169.8, 943)	809 (300, 1365)	216 (85, 640)	0.02
Hypocomplementemia	5%	5%	20%	6%	<0.0001

Conclusions: Using an unbiased method, we identified four phenotypic clusters of IgG4-RD patients. In addition to the differences in organ involvement, clusters were distinguished by age at diagnosis as well as race/ethnicity and gender distribution, serum IgG4 concentrations, and frequency of hypocomplementemia. These clusters may identify patients with IgG4-RD resulting from different risk factors or exposures and those likely to respond differently to treatment.

Disclosure of Interest: None declared

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OP0082 APREMILAST FOR BEHÇET'S SYNDROME: A PHASE III RANDOMISED, PLACEBO-CONTROLLED, DOUBLE-BLIND STUDY (RELIEF)

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Background: Oral ulcers (OU) are the most common sign of Behçet's syndrome (BS) and are observed in nearly every patient. Due to their severity and frequency of recurrence, OU can be disabling and have a substantial effect on quality of life. There is an unmet need for effective treatment for OU in BS. Apremilast (APR), an oral phosphodiesterase 4 inhibitor that modulates inflammatory pathways, demonstrated efficacy in the treatment of oral and genital ulcers of BS in a phase II study.

Objectives: Phase III study to further evaluate the efficacy and safety of APR for OU in BS pts with active OU previously treated with ≥ 1 medication.

Methods: In this phase III, multicenter, randomised, placebo (PBO)-controlled study, 207 eligible pts were randomised (1:1) to APR 30 mg BID (n=104) or PBO (n=103) for 12 weeks, followed by a 52 week active-treatment extension. Pts had active BS, with ≥ 3 OU at randomization or ≥ 2 OU at screening +randomization, without active major organ involvement. Primary endpoint was area under the curve (AUC) for total number of OU over 12 weeks. AUC reflects the change in the number of OU over time, accounting for the clinical characteristic that OU repeatedly remit and recur. Secondary endpoints assessed other measures of OU, including pain, OU resolution (OU-free), maintenance of OU resolution, and time to resolution. Effects on genital ulcers were also assessed. A prespecified hierarchical testing procedure was used for multiplicity adjustment.

Results: AUC for total number of OU over 12 weeks was statistically significantly lower in the APR group compared with the PBO group (table 1). This treatment effect is supported by statistically significant benefits in the APR group compared with PBO for secondary endpoints assessing OU, including pain, OU resolution, maintenance of OU resolution, and time to resolution. A numerically greater proportion of pts achieved resolution of genital ulcers at Week 12 in the APR group compared with PBO.

The incidence of treatment-emergent adverse events (AEs) was comparable between APR and PBO during the PBO-controlled period (78.8% vs 71.8%, respectively). Serious AEs were observed in 3 APR pts (migraine, oral ulcer flare, genital ulcer, arthralgia, soft tissue injury) and 4 PBO pts (diarrhoea, genital and fungal infections, oral ulcer flare, acne, acute febrile neutrophilic dermatosis, erythema multiforme).

Endpoints	P Value	Adjusted Treatment Difference (or HR)	APR vs. PBO (LS Mean, Median Time or %)
AUC for the number of OU from baseline through Week 12	<0.0001	-92.6	129.5 vs. 222.1
Change from baseline in the pain of OU as measured by VAS at Week 12	<0.0001	-24.8	-40.7 vs. -15.9
Proportion of subjects achieving OU resolution by Week 6 who remain OU free for ≥6 additional weeks (maintenance of OU resolution)	<0.0001	25.1%	29.8% vs. 4.9%
Time to OU resolution, during the PBO-Controlled Treatment Phase	<0.0001	HR = 2.400	Median time in weeks: 2.1 vs. 8.1
Proportion of subjects achieving resolution of OU at Week 12	<0.0001	30.6%	52.9% vs. 22.3%
Proportion of subjects achieving resolution of genital ulcers at Week 12 for subjects who had genital ulcers at baseline	0.1100	28.4%	70.6% vs. 41.2%

Conclusions: APR effectively reduced the number and pain of OU, improved time to oral ulcer resolution, and maintained the resolution of OU, the most common manifestation of BS. Favourable treatment effects were also observed for genital ulcer resolution. The safety profile was consistent with the known safety profile of APR.

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OP0083 TEMPORAL TRENDS AND MORTALITY OF HOSPITALISED PATIENTS WITH ADULT ONSET STILL'S DISEASE: A NATIONWIDE ESTIMATE

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Background: There is a dearth of epidemiological studies on Adult Onset Still's Disease (AOSD). Majority of studies are from single centres or are regional. The largest AOSD epidemiological study till date describes 512 patients.¹

Objectives: To describe the demographics, complications, mortality and trends of hospitalised patients with AOSD in United States. Also, to understand the factors associated with mortality in these patients.

Methods: All adult (>18 years) hospitalised patients between 2009 and 2013 from a nationwide inpatient sample (NIS) database were captured. AOSD patients were identified using the ICD-9 code 714.2. Patients also coded for Rheumatoid Arthritis, Lupus, Myositis, Polymyalgia Rheumatica, Ankylosing Spondylitis and Psoriatic Arthritis were excluded. This was done in order to capture patients with strictly AOSD. NIS is the largest all-payer inpatient care database in the United States with approximately 8 million hospitalizations each year. Discharge weights were used to enable nationwide estimates. Descriptive statistics were represented as means/medians for continuous and as frequencies and percentages for categorical variables. A survey weighted logistic regression model was used to describe the associations with in-hospital death.

Results: Between 2009 and 2013, 5,820 AOSD patients were hospitalised (table 1). AOSD patients had a mean age of 53.6 (SE - 0.61) years, 3817 (65.6%) were females. The racial/ethnic distribution showed that 56% white, 15% African American, 11.7% Hispanic and 3% Asian patients were affected. Over the years, the number of white patients that are hospitalised seems to have increased. 37.6% were hospitalised in urban teaching hospitals. 100 (1.7%) of patients developed Macrophage Activating Syndrome (MAS). 66 (1.1%) patients had disseminated intravascular coagulation (DIC) and 25 (0.4%) had thrombotic thrombocytopenic purpura (TTP). Mean length of stay was 6.9 days. There were 154 inpatient deaths in 5 years (mortality 2.6%). The patients who died in the hospital had a higher mean age of 62.4 (SE- 3.1) years, had a higher proportion of Asians

(13.9%) and had increased number of comorbid conditions. Asians had 6.4 times the risk of in-hospital death compared to Whites. The risk for in-hospital death is 30 times higher if a AOSD patient had concurrent DIC.

Abstract OP0083 – Table 1 AOSD characteristics

Variables, n (%) / Mean (SE) when specified	2009 n = 846	2010 n = 995	2011 n = 1434	2012 n = 1280	2013 n = 1265	Total N = 5820
Age in Years, Mean (SE)	53.4 (1.3)	52.3 (1.6)	54.9 (1.1)	52.8 (1.2)	53.3 (1.1)	53.6 (0.6)
Female	520 (61.4%)	645 (64.8%)	892 (62.2%)	570 (68%)	890 (70.4%)	3817 (65.6%)
Race						
White	437 (51.6%)	515 (51.8%)	819 (57.1%)	760 (59.4%)	730 (57.7%)	3261 (56%)
Black	117 (13.9%)	168 (16.9%)	243 (16.9%)	180 (14.3%)	165 (13%)	874 (15%)
Hispanic	96 (11.3%)	120 (12%)	142 (9.9%)	120 (9.4%)	205 (16.2%)	683 (11.7%)
Asian	25 (3%)	47 (4.7%)	29 (2%)	25 (2%)	50 (4%)	175 (3%)
Native American + Others	21 (2.5%)	20 (2%)	79 (5.5%)	80 (6.3%)	35 (2.8%)	236 (4.1%)
Missing values	150 (17.7%)	125 (12.5%)	122 (8.5%)	115 (9%)	80 (6.3%)	591 (10.2%)
Insurance						
Medicare	311 (36.7%)	338 (34%)	589 (41.1%)	525 (41%)	570 (45.1%)	2333 (40.1%)
Medicaid	104 (12.3%)	157 (15.7%)	170 (11.9%)	145 (11.3%)	160 (12.6%)	736 (12.6%)
Private including HMO	351 (41.5%)	373 (37.5%)	474 (33.1%)	475 (37.1%)	375 (29.6%)	2048 (35.2%)
Self-pay	60 (7.1%)	79 (7.9%)	95 (6.6%)	85 (6.6%)	80 (6.3%)	398 (6.8%)
No charge/other	20 (2.4%)	49 (4.9%)	98 (6.8%)	50 (3.9%)	75 (5.9%)	292 (5%)
Length of stay (days), Mean (SE)	6.1 (0.5)	8.7 (1.1)	7 (0.4)	6.6 (0.4)	6.3 (0.4)	6.9 (0.3)
Total Charges in USD, Mean (SE)	\$33,152 (2406)	\$25,501 (3024)	\$29,633 (2450)	\$32,080 (2590)	\$31,367 (2137)	\$30,857 (516)
Median household income						
\$1 - \$3999	205 (24.2%)	276 (27.7%)	294 (20.5%)	340 (26.6%)	315 (24.9%)	1430 (24.6%)
\$3999 - \$4999	217 (25.7%)	222 (22.3%)	253 (17.6%)	255 (19.9%)	315 (24.9%)	1262 (21.7%)
\$4999 - \$5999	166 (19.6%)	275 (27.7%)	370 (25.8%)	325 (25.4%)	305 (24.1%)	1442 (24.8%)
\$6000+	236 (27.9%)	188 (18.9%)	498 (34.7%)	335 (26.2%)	315 (24.9%)	1572 (27%)
Bed size of the hospital						
Small	51 (6%)	78 (7.8%)	123 (8.5%)	175 (13.7%)	140 (11.1%)	566 (9.7%)
Medium	169 (20%)	158 (15.9%)	230 (16.1%)	305 (23.8%)	270 (21.3%)	1133 (19.5%)
Large	341 (40.3%)	384 (38.5%)	462 (32.2%)	640 (50%)	610 (48.2%)	2436 (41.9%)
Missing values	285 (33.7%)	375 (37.7%)	620 (43.2%)	160 (12.5%)	245 (19.4%)	1685 (29%)
Location/teaching status of hospital						
Rural	60 (7.1%)	93 (9.4%)	88 (6.2%)	65 (5.1%)	100 (7.9%)	406 (7%)
Urban non-teaching	233 (27.8%)	240 (24.2%)	379 (26.4%)	345 (27%)	340 (26.9%)	1538 (26.4%)
Urban teaching	267 (31.6%)	286 (28.8%)	347 (24.2%)	710 (55.5%)	580 (45.8%)	2191 (37.5%)
Missing values	285 (33.7%)	375 (37.7%)	620 (43.2%)	160 (12.5%)	245 (19.4%)	1685 (29%)

Table 2: Factors associated with In-Hospital Death

Variable	Odds ratio (95% CI)	p-value
Age	1.03 (0.99 - 1.06)	0.154
Female	1.32 (0.5 - 3.49)	0.579
Race		
White	Reference	
Asian	6.39 (1.77 - 23.1)	0.005
Black, Hispanic and Others	0.84 (0.26 - 2.75)	0.772
Number of Chronic conditions	1.20 (1.05 - 1.37)	0.006
Median Household income		
\$1 - \$3999	Reference	
\$3999 - \$4999	1.96 (0.41 - 9.29)	0.398
\$4999 - \$5999	0.95 (0.16 - 5.67)	0.958
\$6000+	2.57 (0.57 - 11.62)	0.221
Disseminated/Intravascular Coagulation	29.69 (5.5 - 160.4)	<0.001

Conclusions: In hospitalised American AOSD patients, the average age was higher than previously described in cross sectional studies. This may indicate an ageing population with a higher number of comorbidities that justify hospitalisation. More patients were in large or urban teaching hospitals compared to small or rural hospitals. In-hospital death was associated with increased comorbid conditions and was significantly higher among Asians and patients with DIC. To our knowledge, this is the largest epidemiological study of AOSD.

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OP0084 SERUM FIBROBLAST GROWTH FACTOR 2 IS A USEFUL BIOMARKER TO DISTINGUISH ADULT ONSET STILL DISEASE FROM SEPSIS

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Background: The precise cytokine networks in the serum of individuals with adult onset still disease (AOSD) that are associated with its pathogenesis have been unknown. Serum levels of interleukin (IL)-1β, IL-6 and IL-18 have been reported as useful serum biomarkers for diagnosis and disease evaluation among AOSD patients,^{1,2} but these cytokines are also elevated in other inflammatory diseases including severe infection.