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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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.