Conventional synthetic DMARDs were used by 65.0% of RA patients, 54.8% by PsA and 9.8% in ax-SpA and for biologic DMARDs the corresponding numbers were 36.7%, 43.9%, 65.9%, respectively.

Percentage of patients in remission was significantly lower in high pain catastrophizers compared with low pain catastrophizers in RA (34.1% vs 70.3%, $p<0.01$) and ax-SpA patients (17.9% vs 64.5%, $p<0.01$) but not in PsA (10.3% vs 26.7%, $p=0.06$).

In analysis adjusting for age, gender, disease duration and global pain (Tab. 1), PC was independently associated with lower remission rate in RA (OR 0.79 (95% CI 0.63, 0.99), $p=0.04$) and in ax-SpA (OR 0.65 (95% CI 0.46, 0.93), $p=0.02$) but not in PsA (OR 0.83 (95% CI 0.55, 1.25), $p=0.37$).

Conclusion: Our data indicates that high level of patient's PC may reduce likelihood of achieving remission in RA and ax-SpA. Interestingly, this was not shown in PsA patients. Further studies are needed to reveal the importance of PC and its influence on composite disease activity measures used to define disease status e.g. remission.

Table 1. Variables and their associations with remission rates in RA, PsA, ax-SpA in logistic regression analysis.

	RA remission		PsA remission		ax-SpA remission	
	OR (95%CI)	p	OR (95%CI)	p	OR (95%CI)	p
PCS	0.79 (0.63, 0.99)	0.04	0.83 (0.55, 1.25)	0.37	0.65 (0.46, 0.93)	0.02
Age	0.99 (0.97, 1.01)	0.17	1.00 (0.96, 1.04)	0.85	0.98 (0.94, 1.02)	0.27
Gender	0.87 (0.52, 1.45)	0.59	0.37 (0.15, 0.93)	0.03	0.92 (0.40, 2.14)	0.85
Disease duration	1.03 (1.00, 1.05)	0.03	0.95 (0.90, 1.00)	0.06	1.04 (0.99, 1.09)	0.09
Global pain (VAS)	0.96 (0.95, 0.97)	<0.01	0.85 (0.81, 0.89)	<0.01	0.90 (0.88, 0.92)	<0.01

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Infection-related rheumatic and orphan diseases

OP0090

CLASSIFICATION OF PATIENTS WITH RELAPSING POLYCHONDRIITIS BASED ON SOMATIC MUTATIONS IN UBA1

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Background: Somatic mutations in ubiquitin activating enzyme 1 (UBA1) cause a newly defined syndrome known as VEXAS. [1] More than fifty percent of patients currently identified with VEXAS meet diagnostic criteria for relapsing polychondritis (RP).

Objectives: To determine the prevalence VEXAS within a cohort of patients with RP, to compare their clinical, laboratory, and immunologic features and to develop a clinical algorithm to inform genetic screening for VEXAS among patients with RP.

Methods: Exome and targeted sequencing of the UBA1 gene was performed in a prospective observational cohort of patients with RP. Clinical and immunological characteristics of patients with RP were compared based on presence or absence of UBA1 mutations. Random forest was used to derive a clinical algorithm to identify patients with UBA1 mutations. Immune populations were quantified by multipanel flow cytometry. Categorical and continuous variables were compared using the chi square or Kruskal-Wallis test. $P<0.05$ defined statistical significance.

Results: Seven of 92 patients with RP (7.6%) were confirmed to have UBA1 mutations (VEXAS-RP). Six additional patients with VEXAS-RP from other cohorts were included for subsequent analyses. Patients with VEXAS-RP were all male, older at disease onset, and commonly had fever, ear chondritis, skin involvement, deep vein thrombosis, and pulmonary infiltrates. Patients with RP as compared with VEXAS-RP had a significantly higher prevalence of airway chondritis, costochondritis and tenosynovitis/arthritis. (Table). Mortality was significantly greater in VEXAS-RP than RP (27% vs 2% $p=0.01$). Maximum ESR, CRP, and mean corpuscular volume (MCV) values were significantly greater in VEXAS-RP. Absolute monocyte, lymphocyte, and platelet counts were significantly lower in VEXAS-RP. A decision tree based on male sex, MCV>100 fl and Platelet count<200 K/ul classified between VEXAS-RP and RP with 100% sensitivity and 96% specificity.

Conclusion: Mutations in UBA1 are causal for disease in a subset of patients with RP. These patients are defined by disease onset in the fifth decade of life or later, male sex, ear/nose chondritis and hematologic abnormalities. Early identification is important in VEXAS given the associated high mortality rate.

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Table 1. Clinical Characteristics of patients with RP vs VEXAS-RP

	All Patients n=98	RP n=85	VEXAS-RP n=13	p value
Demographic Characteristics				
Race, White n (%)	90 (92)	77 (91)	13 (100)	0.59
Sex, Male n (%)	26 (27)	13 (15)	13 (100)	<0.0001
Age, Symptom onset, years, Median (IQR)	38 (30-47)	37 (28-43)	56 (54-64)	<0.0001
Clinical Symptoms				
Fever n (%)	33 (34)	20 (24)	13 (100)	<0.0001
Ear chondritis n (%)	61 (62)	48 (56)	13 (100)	0.0015
Nose chondritis n (%)	83 (85)	71 (84)	12 (92)	0.68
Airway chondritis n (%)	37 (38)	37 (44)	0 (0)	0.0015
Tenosynovitis/arthritis n (%)	83 (85)	77 (91)	6 (46)	0.0005
Skin involvement n (%)	33 (34)	22 (26)	11 (85)	<0.0001
Laboratory Values				
ESR, mm/hr, median (IQR)	12 (6-22)	11 (5-19)	66.5 (42-110)	<0.0001
CRP, mg/L, median (IQR)	2.9 (0.8-9.6)	1.9 (0.6-6.3)	17.7 (9.6-99.5)	<0.0001
Platelet count (k/uL)	246(201-299)	258 (227-312)	145 (100-169)	<0.0001
MCV fL	93.05 (90-98)	92.2 (89-95)	105 (102-115)	<0.0001
Absolute lymphocyte count	1.6 (1.1-2.3)	1.78(1.4-2.4)	0.92 (0.5-1.2)	<0.0001
CT scan abnormalities				
Pulmonary infiltrates n (%)	16 (16.33)	6 (7.06)	10 (77)	<0.0001
Complications				
Death n (%)	6 (6)	3 (4)	3 (23)	0.029
Unprovoked DVT	12 (12)	4 (5)	8 (62)	<0.0001

N number; IQR = interquartile range

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