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Background: Behçet's Syndrome (BS) is a vasculitis of unknown origin. Vascular involvement, so-called vascular Behçet's syndrome (VBS), may involve blood vessels of all sizes belonging to both venous and arterial system, with pulmonary artery involvement (PAI) being the most frequent form of arterial involvement. PAI in BS occurs in the form of aneurysms or thrombosis, and results in significant mortality.

Objectives: To report the clinical characteristics of PAI in patients with BS and to define the predictors of relapses.

Methods: We performed a retrospective analysis of BS patients with PAI who fulfilled international study group criteria. Among 460 patients with VBS, 66 were diagnosed with PAI. For final analyses, 61 patients with PAI, who had at least 2 follow-up visits (72.1% male, mean age at BS diagnosis 29.34 (SD 10.1) years), were included. The data of the patients were recorded. Relapse was defined as the reoccurrence of vascular event at any vascular structure. Factors associated with relapse were assessed by logistic regression analysis.

Results: There were not any differences considering demographic and clinical features of the patients with and without PAI in VBS group except for that intracardiac thrombosis was more common in the patients with PAI than the patients without (19.7% vs 0.3%, respectively) (Figure 1). Among 61 patients with PAI, 50 (82.0%) had isolated pulmonary artery thrombosis (PAT), whereas 11 (18.0%) had pulmonary artery aneurysm (PAA) with or without PAT. The characteristics of 61 patients with PAI were shown in Table 1. A total of 37 relapses occurred in 24 (39.3%) patients during follow-up a median of 65.9 (IQR 20.1-109.0) months. To define the factors associated with relapses, the patients with isolated PAT were analysed. In multivariable logistic regression analysis, older age at BS diagnosis and anticoagulation usage seemed to be protective, even though they could not reach statistical significance (OR: 0.92 95% CI 0.86-1.02, OR: 0.34 95% CI 0.09-1.33, respectively).

Figure 1. A) Axial CT image shows thrombus formation in the left upper lobe pulmonary artery branch. B) Giant thrombus is seen in the right ventricle (black arrow). Note the thrombus formation in the left lower lobe pulmonary artery branches (white arrows).

Table 1. Characteristics of the patients with pulmonary artery involvement

	All n= 61	Isolated PAT n= 50	PAA n= 11
Sex (male), n (%)	44 (72.1)	35 (70.0)	9 (81.8)
Age at diagnosis of BS, years (SD)	29.3 (10.1)	28.8 (10.4)	31.9 (8.0)
Age at diagnosis of PAI, years (SD)	36.3 (12.83)	36.7 (13.8)	34.7 (6.7)
Symptoms, n (%)			
Dyspnea	28 (45.9)	23 (46.0)	5 (45.5)
Hemoptysis	26 (42.6)	19 (38.0)	7 (63.6)
Vascular event before PAI, n (%)	21 (34.4)	20 (40.0)	1 (9.1)
Other vascular events at PAI, n (%)	33 (54.1)	28 (56.0)	5 (45.5)
Thrombophlebitis, n (%)	7 (21.2)	6 (21.4)	1 (20.0)
Upper and/or lower extremity DVT, n (%)	20 (60.6)	16 (57.1)	4 (80.0)
SVCS, n (%)	1 (3.0)	1 (3.6)	0 (0)
IVCS, n (%)	3 (9.1)	1 (3.6)	2 (40.0)
Intracranial thrombosis, n (%)	4 (12.1)	4 (14.3)	0 (0)
Intracardiac thrombosis, n (%)	8 (24.2)	7 (25.0)	1 (20.0)
Arterial event other than PAI, n (%)	4 (12.1)	3 (10.7)	1 (20.0)
Relapse, n (%)	24 (39.3)	20 (40.0)	4 (36.4)
Time to the first relapse, median (IQR), month	49.8 (13.2-102.6)	54.1 (24.4-117.9)	3.9 (1.7-67.9)
Death, n (%)	2 (3.3)	0 (0)	2 (18.2)

BS= Behçet Syndrome, PAI= pulmonary artery involvement, PAT= pulmonary artery thrombosis, PAA= pulmonary artery aneurysm, DVT= deep vein thrombosis, SVCS= superior vena cava syndrome, IVCS= inferior vena cava syndrome

Conclusion: Our results indicate that there is a higher frequency of intracardiac thrombosis in BS patients with PAI and an increasing current trend of PAT in patients with PAI. In addition, there might be a possible efficacy of anticoagulation usage in preventing relapses, which needs confirmation with further studies.

Disclosure of Interests: None declared

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POS1336

RETROPERITONEAL FIBROSIS IN ERDHEIM-CHESTER DISEASE HAS UNIQUE PRESENTING AND PROGNOSTIC FEATURES: A SINGLE CENTRE RETROSPECTIVE COMPARATIVE COHORT STUDY

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Background: Erdheim-Chester disease (ECD) is a multisystemic non-Langerhans cell histiocytosis. Retroperitoneal fibrosis (RPF) is a common ECD feature, found in up to 60% of patients. Besides ECD, RPF differential diagnosis includes other clinical entities of rheumatological interest, such as idiopathic RPF and IgG4-related disease (IgG4-RD). Since ECD management does not include glucocorticoids and mainly relies on the use of anti-neoplastic small molecules, a prompt diagnosis is critical.

Objectives: To characterize the main differential clinical, radiologic and prognostic features of a monocentric cohort of patients with RPF. To provide practical learning points useful to recognize ECD among patients with RPF.

Methods: Medical records of ECD patients with radiologic signs of RPF and of patients with non ECD-related RPF followed up at our Rheumatology Clinic were retrospectively reviewed. Final diagnosis relied either on analysis of histologic samples or on fulfilment of appropriate classification criteria. Main clinical, radiologic and laboratory features at diagnosis and long-term outcomes were evaluated and compared between the two groups. Non parametric tests were used for comparison.

Results: 31 (41%) ECD and 45 (59%) non-ECD patients (IgG4-RD, n=28; idiopathic RPF, n=11; peri-aortitis with large-vessel involvement, n=6) were included. All but one (97%) ECD patients and 56% non-ECD patients had a biopsy-proven diagnosis. ECD patients were younger at disease onset (51±14 vs 59±10 years, P=0.024) and diagnosed with a greater delay (48 [IQR, 13-92] vs 4 [IQR, 2-7] months, P<0.001). In the majority of ECD patients (84%), RPF was clinically silent and was disclosed in the context of the diagnostic work-up; conversely, 82% non-ECD patients had at least one RPF-related symptom (mainly, abdominal pain, n=19, and low-back pain, n=16). At diagnosis, C-reactive protein (50±34 vs 33±31 mg/L, P=0.052) and serum creatinine (1.31±0.53 vs 1.34±0.79, P=0.76) levels were not statistically different. In all ECD patients, fibrotic tissue surrounded both kidneys symmetrically, involving the ureters in half of the cases (Table 1); signs of hydronephrosis were present in 55% of patients (mostly bilaterally, 77%). In the non-ECD group, kidneys were rarely affected, and ureteral involvement was mostly asymmetric; hence, hydronephrosis incidence was similar (76%), but more frequently monolateral (65%). In non-ECD patients, fibrotic tissue more frequently involved inferior vena cava and iliac arteries. FDG uptake by RPF was disclosed in 39/40 (98%) non-ECD and 5/12 (42%) ECD patients who underwent a PET before therapy start (P<0.001). Ureteral stenting was required by a similar fraction of patients (ECD, 29%; non-ECD, 38%, P=0.47), but it was bilateral in 100% of ECD vs 29% of non-ECD patients (P<0.001). No differences in the rate of chronic kidney failure was found (26% vs 18%, P=0.41), but only ECD patients progressed to a terminal stage requiring hemodialysis (13% vs 0%, P=0.025).

Table 1.

	ECD patients (n=31)	Non-ECD patients (n=45)	P-value
Left kidney	30 (97%)	4 (9%)	<0.001
Right kidney	31 (100%)	6 (13%)	<0.001
Left ureter	16 (52%)	16 (36%)	0.24
Right ureter	16 (52%)	24 (53%)	1
Abdominal aorta	21 (68%)	40 (89%)	0.04
Inferior vena cava	5 (16%)	29 (64%)	<0.001
Left iliac artery	10 (32%)	27 (60%)	0.02
Right iliac artery	10 (32%)	29 (64%)	0.001
Left renal artery	9 (29%)	6 (13%)	0.14
Right renal artery	6 (19%)	4 (9%)	0.3
Mesenteric artery	8 (26%)	4 (9%)	0.06
Celiac trunk	7 (23%)	3 (7%)	0.08

Conclusion: Our retrospective study suggests that ECD-related RPF has a significantly worse renal prognosis than non ECD-related RPF. A complete baseline work-up including a critical evaluation of the extension of the fibrotic tissue and the sites involved is fundamental to avoid patients' misdiagnosis and mistreatment.

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POS1337 **BEHÇET'S DISEASE AND COURSE OF PREGNANCY - RESULTS FROM THE NATIONWIDE PREGNANCY REGISTER REVNATUS**

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Background: Behçet's disease (BD) is a complex and multisystemic disorder of unknown etiology. It is characterized by recurrent oral and genital ulcers and inflammatory eye involvement. It can also affect the cardiovascular, pulmonary and skeletal system. BD is commonly diagnosed in women in reproductive age. Pregnancy outcomes are reported in previous studies with discrepancies in the rate of pregnancy-related complications.

Objectives: To describe the course of pregnancy in patients with BD.

Methods: We prospectively collected data on pregnancy outcome in women with BD through the nationwide Norwegian registry on pregnancy and rheumatic diseases (RevNatus) from 2016-2020. All women were diagnosed with ICD-10 code M35.2. 13 pregnancies in 11 women were recorded. One woman had two pregnancies including one twin birth.

Results: Median age at conception was 30 years (range 25-32), with median disease duration 7 years (range 1-15). There were no serious organ manifestations. Tnf-alpha-inhibitors were used in 2nd trimester in 3 pregnancies. All pregnancies resulted in live births with median gestational age at delivery of 38 weeks (range 36-41). No preeclampsia or HELLP were recorded.

Table 1. Pregnancy outcome in 13 BD pregnancies

Patient #	Pregnancy #	Medication at 2.nd trimester	Oral or genital ulcers during pregnancy	Disease activity during pregnancy	CRP value at 2nd trimester	Gestational age at delivery, week	Mode of delivery	Birth weight, gram
A	1	Azathioprine Colchicine LMWH	no	persistent*	13	38	SVD	3475
B	2	Adalimumab Colchicine LDPrednisolone	yes	persistent *	4	40	SVD	3215
C	3	no	yes	persistent*	5	41	SVD	3355
D	4	Adalimumab LDPrednisolone	yes	persistent*	23	36	SVD	1940
E	5	Etanercept ASA	no		2	41	SVD	3410
	6a (twin)	Etanercept ASA	no		5	37	Elective Sectio	2400
	6b (twin)							2410
F	7	LDPrednisolone	yes	flare**	15	38	SVD	3180
	8	Adalimumab LDPrednisolone	yes	flare**	4	37	Acute Sectio	3049
G	9	no	no	remission***	1	41	SVD	3150
H	10	LDPrednisolone	yes	persistent *	10	38	SVD	2480
I	11	ASA	no	persistent*	3	39	SVD	3410
J	12	no	yes	persistent*	1	37	SVD	3115
K	13	Ciklosporine	no	remission***	3	37	SVD	3130

SVD = spontaneous vaginal delivery, ASA = acetylsalicylic acid, LDPrednisolone = Low dose Prednisolone <10 mg/day, * = persistent throughout pregnancy, ** = flare once during pregnancy, *** = remission throughout pregnancy

Conclusion: Pregnancies in women with BD in this cohort were uncomplicated. We did not reveal any serious events, and they all resulted in live births. However, BD is a very heterogeneous disease, and the women in our cohort did not have any serious organ manifestations. Larger cohorts representing the heterogeneity are needed to study the course and outcome of pregnancies in BD.

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POS1338 **INCIDENCE, PREVALENCE, AND MORTALITY OF CHRONIC PERIAORTITIS: A POPULATION-BASED STUDY**

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Background: Chronic periaortitis is an inflammatory condition that typically involves the infrarenal portion of the abdominal aorta. Few studies are available to outline the epidemiology of this rare condition. To date, no epidemiologic studies on periaortitis have been performed in North America.

Objectives: To evaluate the epidemiology, presentation and outcomes of patients with chronic periaortitis from 1998 through 2018.

Methods: An inception cohort of patients with incident chronic periaortitis from January 1, 1998 through December 31, 2018, in Olmsted County, Minnesota, USA, was identified based on comprehensive individual medical record review utilizing the Rochester Epidemiology Project medical record linkage system. Inclusion required radiographic and/or histologic confirmation of periaortic soft tissue thickening around at least part of the infra-renal abdominal aorta or the common iliac arteries. Data were collected on demographic characteristics, clinical presentation, renal and radiographic outcomes, and mortality. Incidence rates were age and sex adjusted to the 2010 United States white population.

Results: Eleven incident cases of chronic periaortitis were identified during the study period. Mean±SD age at diagnosis was 61.8±13.4 years. The cohort included 9 men (82%) and 2 women (18%). The most common presenting symptom was pain with 55% (6/11) reporting abdominopelvic-pelvic pain, 36% (4/11) back pain, and 18% (2/11) flank pain. Obstructive uropathy was present in 73% (8/11) subjects: 3 (27%) unilateral left, 1 (9%) unilateral right, and 4 (36%) bilateral. Mean creatinine at presentation was 2.7±3.4 mg/dL. Ureteral stenting was required at diagnosis in seven patients: unilateral left in 2, unilateral right in 1 and bilateral in 4. All 11 patients received glucocorticoids with a median (IQR) dose of 40 (30, 60) mg/day. Additional non-glucocorticoid therapeutics were used in 10 patients.

Renal function stage at last follow up declined in 2 patients, remained the same in 3 patients and improved in 6 patients. Mean creatinine at last follow-up was 1.2±0.2 mg/dL. Among the seven patients requiring baseline indwelling ureteral stent placement only two required ongoing ureteral stenting at last follow up. None of the four patients without ureteral stenting at diagnosis progressed to require stenting during the follow-up period. No patient underwent ureterolysis surgery in this cohort. Periaortic soft tissue thickening at last follow up had increased in thickness in 1 (9%), was unchanged in 2 (18%), decreased in size but did not resolve in 6 (55%), and fully resolved in 18%.

Age- and sex-adjusted incidence rates per 100,000 population were 0.26 for females, 1.56 for males and 0.87 overall. Overall prevalence on January 1, 2015 was 8.98 per 100,000 population. Median (IQR) length of follow-up was 10.1 (2.5, 13.8) years. Overall mortality was similar to the expected age, sex, and calendar estimates of the Minnesota population with standardized mortality ratio (95% CI) for the entire cohort 2.07 (0.67, 4.84).

Conclusion: This study reports the first epidemiologic data on chronic periaortitis in the United States. In this cohort of patients with chronic periaortitis, men were approximately 4 times more commonly affected than women. Mortality was not increased compared to the general population.

Disclosure of Interests: None declared

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POS1339 **MORE FREQUENT AND EARLIER HIP INVOLVEMENT IN SPONDYLOARTHRITIS ASSOCIATED WITH FAMILIAL MEDITERRANEAN FEVER**

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Background: Familial Mediterranean fever (FMF) is a hereditary autoinflammatory disorder caused by the MEFV gene variants. Although association between FMF and spondyloarthritis (SpA) has previously been reported, clinical and laboratory features of patients with FMF and SpA have not been defined in detail.

Objectives: We aimed to evaluate clinical and laboratory characteristics, disease outcome and biologic responses of patients with FMF+SpA compared to patients with only SpA who were followed-up in our tertiary referral center.

Methods: Database of FMF Clinic was screened for FMF patients with coexistent SpA and 113 patients were identified fulfilling Tel Hashomer and ASAS criteria for FMF and SpA, respectively. A group of patients with SpA without FMF matched for age, gender and disease duration were selected as the control group.

Results: Thirteen patients were excluded because of missing data, and 100 patients (F/M: 52/48) were included into the analysis. Mean follow-up time was 93.6 ± 77 (range [r]: 3-324) months and mean patient age was 43.3 ± 12 (r: 20-87) years. Mean age of onset for FMF was 12.5 ± 8 (r: 1-36) and for SpA was 25 ± 11 (r: 7-72) years. SpA findings was classified as axial in 35.4%, axial and peripheral in 47.9% and only peripheral in 16.7% in FMF+SpA group. Half (49%) of the