Table 1. *ACR / EULAR 2017 provisional classification criteria for GPA **ACR 1990 Classification criteria for EGPA ***Modified diagnostic criteria for Takayasu arteritis. Shanna et al. 1995. Abbreviations: AZA: Azathioprine; CYC: Cyclophosphamide; EGPA: Eosinophilic Granulomatosis with Polyangiitis; ENG: electroneurography; GPA: Granulomatosis with Polyangiitis; IgG4RD: IgG4 Related Disease; INFX: Infliximab; F: female; M: male; MPDN: Methylprednisolone; MTX: Methotrexate; N: No; PDN: Prednisone; RTX: Rituximab; TCZ: Tocilizumab; Y: Yes

GENDER (Age at IgG4RD diagnosis^)		M (56)	M (69)	F (47)	F (51)	M (65)	F (16)
IgG4RD	Organs involved	pancreas, biliary tract, lymphnodes, nasal cavities, skin, ascending aorta	salivary glands, pancreas, biliary tract, lung	eylid	periorbital region, lacrimal gland	meninges, abdominal aorta	peritoneum
	ACR/EULAR score	34	36	20	21	24	26
	Definite histological diagnosis	Y (pancreas, lymphnodes)	Y (salivar glands, lung)	Y (eyelid)	Y (periorbiatl tissue, lacrimal gland)	Y (meninges)	Y (peritoneal lesion?)
	Induction of remission therapy	PDN + AZA	PDN	AZA	PDN	PDN + MTX	PDN + MTX
	Response	Y	Y	Y	Y	Y	Y
	Relapse (years after treatment)	Y (6)	Y (10)	N	Y (0,5)	Y (5)	N
	Second line therapy	RTX	RTX	-	MPDN + RTX	PDN	-
OTHER IMMUNOLOGICAL DISEASE	Diagnosis	Sarcoidosis	Sarcoidosis	GPA	GPA	EGPA	Takayasu
	Organs involved	lymphnodes over and infra diaphragmatic, axillary, laterocervical, lumboaortic, inguinal, hepatic	pulmonary and over and infra-diafragmatic lymphnodes	tracheal cartilage	pulmonary nodules, scleritis	respiratory tract (asthma), skin, peripheral nervous system, blood eosinophilia	left subclavian artery, left common carotid, anonymous artery, thoracic aorta, suprarenal abdominal aorta, right pulmonary artery
	Classification criteria fulfilled?	not available	not available	Y*	Y*	Y**	Y***
	Autoantibody	none	none	c-ANCA	c-ANCA	p-ANCA	none
	Definite histological diagnosis	Y (lymphnode)	Y (paratracheal lymphnodes)	not available	Y (pulmonary nodule)	Y (skin biopsy)	not available
	Timing of diagnosis vs IgG4RD	subsequent	previous	previous	contemporary	previous	contemporary
	Years after/before IgG4RD diagnosis	2	1	12	0	3	0
	Treatment	PDN	PDN + MTX	PDN + AZA	PDN	PDN + CYC	INFX
	Response to treatment	Y	Y	Y	Y	Y	N

AB1295

TREATMENT OUTCOMES OF ADULT-ONSET STILL'S DISEASE PATIENTS WITH BIOLOGICAL THERAPY

Y. Erez[1], T. Demirci Yildirim[2], M. Birlik[2], G. Can[2], F. Onen[2], İ. Sari[2]. [1]*Dokuz Eylul University School of Medicine, Division of Rheumatology, İzmir, Turkey*; [1]*Dokuz Eylul University School of Medicine, Division of Rheumatology, İzmir, Turkey*

Background: Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disorder with heterogeneous distrubition (1). Due to rarity of AOSD and limited controlled study about efficacy of biologic treatment in AOSD, management of refractory AOSD patients poses several challenges.

Objectives: The aim of this study to investigate clinical characteristics and treatment outcomes of AOSD patients who receive biological therapy.

Methods: Patients were identified who met Yamaguchi classification criteria between January 2010 and December 2021 from our database. AOSD patients treated with biologic treatment were included in this study. Demographic data, clinical features and treatment characteristics were recorded.

Results: 49 AOSD patients were identified from database. 9 AOSD patients with biologic therapy (7 female, mean age 42.3±19) were included in this study. Mean age at diagnosis was 31.5± 16.7 and mean follow-up period was 9.3± 5.5 years. Median drug retention duration was found 95 (5-150) months. It was established that all patients used high dose steroid at diagnosis and 3 (33.3%) out of them received pulse steroid therapy. Initial therapy was methotrexate for all patients and leflunomid was used secondly as monotherapy or combination therapy after methotrexate inefficacy. It was observed that indication for biologic therapy was arthritis and systemic inflammatuar symptoms in 5 patients. 4 patients was initiated biologic therapy for refractory arthritis without systemic inflammation findings. First biologic agents were tocilizumab and etanercept in 3 patients, infliximab and anakinra in 2 patients. It was shown that biological therapy was switched in 3 patients due to inefficacy and 2 patients because of side effects. Allergic reaction developed in 2 patients treated with infliximab and infliximab switched to adalimumab and etanercept. Biological therapy was discontinued in 2 patients due to remission, and patients remained in remission after cessation of biological agent. None of patients had serious infection. Hemophagocytic syndrome was developed in only one patient during disease course.

Conclusion: According to our cohort, one of five AOSD patients received biological therapy during follow-up. Our results revealed that the need for biologic therapy during the course of the disease is not uncommon in AOSD, and that biologic therapy is essential for maintaining remission in resistant patients.

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Table 1. Baseline clinical and treatment characteristics (n=9)

Female, n (%)	7 (77.7)
Age, years	42.3±19
Age at diagnosis, years	31.5± 16.7
Smokers, n (%)	6 (66.6)
Drug retantion duration, months	95 (5-150)
Follow-up time, years	9.3± 5.5
Treatment characteristic	n (%)
Biologic therapy indication	
-Systemic inflammation findings	5 (55.5)
-Refractory arthritis	4 (44.4)
İnitial biologic therapy	
Etanercept	3 (33.3)
Tocilizumab	3 (33.3)
Infliximab	2 (22.2)
Anakinra	2 (22.2)
Switching agent	5 (55.5)
Cessation of biologic therapy	3 (33.3)

Disclosure of Interests: None declared
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AB1296

LONG-TERM EXPERIENCE OF THE THERAPY WITH INTERLEUKIN-1 INHIBITORS IN MONOGENIC AUTOINFLAMMATORY SYNDROMES AND SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS IN REAL-LIFE PRACTICE OF RHEUMATOLOGISTS IN SINGLE FEDERAL CENTER

S. Salugina[1], M. Kaleda[1], E. Fedorov[1], I. Nikishina[1], A. Torgashina[2]. [1]*V.A.Nasonova Research Institute of Rheumatology, Pediatric Department, Moscow, Russian Federation*; [2]*V.A.Nasonova Research Institute of Rheumatology, Laboratory of Intensive Therapy Methods, Moscow, Russian Federation*

Background: Autoinflammatory diseases (AIDs) are a heterogeneous group of rare genetically determined conditions. Biologics (B) are used, especially inhibitors of interleukin 1 (iIL-1), in the treatment of the most common monogenic AIDs (mAID) – FMF, TRAPS, HIDS/MKD, CAPS. IIL-1 has shown high efficiency and led to significant progress in the treatment of these patients (pts). Currently, iIL1 are the first-line drugs for mAIDs therapy, primarily CAPS. In pts with systemic juvenile idiopathic arthritis (sJIA) IL-1 inhibitors are also successfully used in the first or second line of therapy along with IL-6 inhibitors.

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