Maintenance therapy. Four patients died during follow-up. table 2 shows data from other worldwide region compared with our data.

Abstract AB0691 – Table 1. Characteristic of chilean patient with ILD an MPA

Age	Sex	CT	Onset (month)	RENAL	MUCOCUTANEOUS	NEUROLOGIC	MSQ	TREATMENT	MORTALITY
55	M	UIP	C	NO	NO	MM	MP/51	CS/CJ/AZA	NO
68	F	UIP	B (10)	NO	LR	PNP	MP/TJ	CS/CJ/AZA	NO
79	M	UIP	B (5)	NO	LR	PNP	MP/51	CS/CJ/AZA	NO
69	F	UIP	C	NO	LR	PNP	SI	CS/CJ/AZA	NO
77	F	UIP	C	GN	LR / CV	MM/NSHL	SI	CS/CJ/AZA	YES
64	F	UIP	C	GN	NO	NO	MP/TJ	CS/CJ/AZA	NO
81	M	UIP	C	GN	NO	NO	NO	CS/CJ/AZA	NO
84	F	UIP/DAH	C	RPGN	Oral Ulcers	NO	NO	CS/CJ/AZA	YES
63	F	UIP	B (168)	GN	NO	CHS/NSHL	NO	CS/CJ/AZA	NO
63	M	NSIP	B (24)	NO	NO	MM	SI	CS/CJ/AZA	NO
53	F	NSIP	C	GN	LR	NO	TJ	CS/CJ/RTX/AZA	YES
68	F	NSIP/DAH	B (50)	NO	NO	PNP	MP	CS/CJ/PLX/AZA	YES
57	M	NSIP/DAH	C	RPGN	NO	PNP	NO	CS/CJ/AZA	NO
78	M	Not UIP not	C	NO	NO	NSHL	MP/51	CS/CJ/AZA	NO
54	F	NSIP	C	NO	LR	PNP	TJ	CS/CJ/AZA	?
32	F	UIP/DAH	B (84)	NO	CV	NO	NO	CS/CJ/AZA	NO

Abstract AB0691 – Table 2. Characteristic of patient with ild and MPA worldwide

Country	CHI	ARG	MEX	CHN	JAP	USA	CAN	PER	IRA
Number of Patient	16	9	19	19	19	3	6	10	40
Age (average)	65.3	58.4	54.2	63.6	66.2	72.3	69.8	70	68
Sex (% female)	62.5	55.5	47.1	58	48	76.7	50	20	39
Onset (M/C/A) ILD-MPA	37.5/62.5/0	55.5/54.5/0	82.3/17.7/0	88.4/11.5/0	52.6/47.3/0	33.3/66.7/0	50/50/0	50/50/0	45/45/12
TC (% UIP/NSIP/Non UIP NSIP)	62.5/37.5/0/0	88.8/11.2/0	100/0/0	100/0/0	100/0/0	0/0/100	100/0/0	60/40/0	NA
Mortality %	25	44.4	41.2	31	47	33.3	83.3	50	36

Conclusions: Among chilean patients there are more females, have a more NSIP pattern, and less mortality that other worldwide series.

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AB0692 BELIMUMAB IN COMBINATION WITH AZATHIOPRINE FOR REMISSION MAINTENANCE IN GRANULOMATOSIS WITH POLYANGIITIS AND MICROSCOPIC POLYANGIITIS: EFFECT ON BIOMARKERS

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Background: GPA and MPA (related types of ANCA-associated vasculitis [AAV]) are organ-/life-threatening systemic vasculitides. B cells and increased circulating BLYS (a B cell survival factor) are implicated in AAV pathogenesis, suggesting a role for BLYS in these vasculitides. BEL is an anti-BLYS human immunoglobulin (Ig) G1 α monoclonal antibody.
Objectives: Examine the effects of BEL plus azathioprine (AZA), on biomarkers in patients with AAV for remission maintenance following standard induction (IND) with glucocorticoids (GC) and CYC or RTX.
Methods: This double-blind, placebo-controlled, multicentre study (BEL115466/ NCT01663623) randomised (1:1) patients (≥ 18 years) with new/relapsing AAV following IND (oral or IV CYC or RTX), to AZA 2 mg/kg/day and oral GC, plus IV BEL 10 mg/kg or PBO (Days 0, 14, 28 and every 28 days until completion). Remission was defined as Birmingham Vasculitis Activity Score=0, plus GC ≤ 10 mg/day. The study was truncated after initiation (n~300 to~100) due to revised AAV standard of care (SoC) affecting recruitment. Biomarker endpoints (serum Igs, B cells and ANCA [anti-MPO/PR3]) were measured at baseline (BL) and thereafter. Summaries by IR were post hoc; no analyses were performed.

Abstract AB0692 – Table 1. Baseline disease characteristics

n (%)	BEL (n=53)	PBO (n=52)
GPA	42 (79)	41 (79)
MPA	11 (21)	11 (21)
ANCA type (historical diagnosis)		
Anti-PR3	41 (77)	40 (77)
Anti-MPO	12 (23)	12 (23)
ANCA positive*	24 (49)	22 (44)
IR		
IV CYC	21 (40)	24 (46)
Oral CYC	18 (34)	15 (29)
Rituximab	14 (26)	13 (25)

ANCA, anti-neutrophil cytoplasm antibodies; BEL, belimumab; CYC, cyclophosphamide; GPA, granulomatosis with polyangiitis; IR, induction regimen; IV, intravenous; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PBO, placebo; PR3, proteinase 3; RTX, rituximab
*n=49 (BEL); n=50 (PBO)

Results: At BL, RTX patients had B cell counts $\leq$ LLQ. CYC patients had notably low BL B cell counts: the lowest were in oral-CYC patients. Circulating memory B cells (CD20+/CD27+) increased rapidly with BEL, then gradually returned to BL (CYC); no major changes occurred with PBO. BEL had no impact on the proportion of naive CD20+CD27– B cells vs PBO, post CYC IND. The number of RTX patients with quantifiable data was low; partial reconstitution occurred in a minority of patients (PBO 2/13; BEL 4/14) and did not translate into vasculitis relapses. Overall Ig levels declined more noticeably with BEL vs PBO; suggesting BEL affects antibody-secreting cells. ANCA+patients were similar between groups (PBO 32/50; BEL 30/49). No trends in change in ANCA status over time occurred, regardless of IR. Individual patient data showed no apparent trends between ANCA titres and AAV activity.

Conclusions: Choice of IR for active AAV affects B cell dynamics. BEL pharmacodynamic effects occurred in patients with AAV receiving SoC. Given the small sample size and high variability, data must be interpreted with caution.

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AB0693 DEMOGRAPHIC, CLINICAL, LABORATORY AND IMAGING CHARACTERISTICS OF AN INCIDENCE COHORT OF 93 PATIENTS WITH LARGE VESSEL GCA

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Background: Few study have evaluated the clinical and imaging data of patients with large vessel GCA (LVGCA)
Objectives: To evaluate clinical, demographical and imaging data of an inception cohort of 93 patients with LVGCA recruited in Reggio Emilia (Northern Italy)
Methods: All patients with incident large vessel GCA diagnosed between 1 January 2005 and 31 December 2016 and living in the Reggio Emilia area, were identified by capture and re-capture checking of computerised discharge diagnosis codes (ICD10) and using outpatients databases from rheumatology, internal medicine, surgery, pathology, imaging departments of Reggio Emilia Hospital as well

as by examining the Reggio Emilia district database for rare diseases. To be included in the study, patients must satisfy the following 2 criteria: Age at disease onset ≥ 50 years; evidence of large-vessel vasculitis by clinical criteria, angiography, MRA, CTA, PET/CT and/or ultrasonography. Demographic, clinical, laboratory and imaging data collected at first visit were retrieved from patients records.

Results: There were 93 incident cases of LV GCA (66 women, 71%) during the 12 year study period; Mean $\pm$ SD age at diagnosis was 72 $\pm$ 9 years. The three most prevalent signs or symptoms were: systemic in 49 pts (52.7%), GCA cranial symptoms in 39 pts (41.9%) and PMR in 35 pts (37.6%). Peripheral ischaemic symptoms were observed only in 8 pts (8.6%). Forty four pts had temporal artery biopsy and 70.5% resulted positive. At US examination the three most common involved arteries were: common carotid artery (59.2% pts), subclavian artery (53.3% pts) and axillary artery (49.3 pts). At PET/CT scan examination the most common involved arteries were: thoracic aorta (72.1% pts), subclavian artery (61.8% pts) and abdominal aorta (58.8% pts). Celiac trunk and mesenteric artery were involved in 18.4% of pts and renal artery in 10.2% of pts.

Conclusions: In an inception cohort of LVGCA systemic manifestations had the highest prevalence among presenting symptoms. Imaging studies demonstrated an high prevalence of aortic and subclavian involvement.

Disclosure of Interest: None declared

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AB0694 GIANT CELL ARTERITIS EPIDEMIOLOGY IN LA RÉUNION: A RETROSPECTIVE CASES SERIES

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Background: Giant Cell Arteritis (GCA) is the most common vasculitis in people over 50 years, with incidence varying according to geographic location. In Europe, the average incidence is 10–30 per 1 000 000 inhabitants over 50 years, whereas in Africa and Asia it is approximately 1.5 per 1 000 000.

Objectives: To define GCA epidemiology in La Réunion, a French overseas territory in South West Indian Ocean characterised by several ethnic groups and genetic admixture.

Methods: Retrospective study between January 2005 and August 2017 in the 4 main hospitals of La Réunion. Patients were identified through hospital informatics databases and practitioner records. A definite diagnosis of GCA was considered for patients fulfilling ACR criteria and/or by assessment by a trained rheumatologist. Incidence and prevalence calculation was based on French national census data.

Results: Sixty patients were included, of which 60% were women. Mean age at diagnosis was 73.7 years, with a mean delay to diagnosis of 6.8 months. Cases met an average of 3.78/5 ACR criteria, and 78% had >2 criteria. The mean annual incidence was 2.33 per 1 000 000 inhabitants older than 50 years (203,000), with 95% confidence interval (IC) of 1.74–2.92. The prevalence at the end of period study was 24 cases per 100,00 (IC: 18–30). There was no seasonal variation regarding disease onset. Clinically, patients complained of asthenia and headache in 75%, fever in 33% and ophthalmologic damage for 32% of the cases, of which 5 had anterior ischaemic optic neuropathy. Polymyalgia rheumatica was associated in 42% of all cases. Total blood cells counts were usually within normal values, whereas mean CRP was 111 mg/L. Anicteric cholestasis was a common finding: mean GGT=78 IU/L (n=42) and alkaline phosphatase=107 IU/L (n=104). Radiological examinations contributed to diagnosis in only 9/31 cases. Temporal artery biopsy was performed in 91% of patients and showed specific histological features of GCA in 55%. Corticosteroid regimen was introduced in 59 patients (1 died the day after the biopsy) and mean treatment duration was 26 months, for a total dose of 10.6 g. Antiplatelet therapy was given in 47% of patients. One patient in three experienced one or more relapses.

Conclusions: This is the first study to describe GCA in La Réunion, and more generally in Indian Ocean. It shows an incidence 4–12 times lower than in most European countries with white ancestry background. This discrepancy could be explained by the contribution of the various ethnic groups of La Réunion, especially those coming from parts of the world characterised by a lower GCA incidence (Africa, India, South East Asia). A shorter life expectancy may also account for this observation, assuming that GCA incidence increases with age. Sex ratio and age at diagnosis were similar to European studies, as well as clinico-biological features, response to treatment and side effects. Some limitations of our study should be taken in consideration: inclusions of hospitalised patients only, informatics record limits and retrospective design that did not afford for ethnic background determination.

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AB0695 PREDICTIVE FACTORS OF LONG-TERM CLINICAL OUTCOME IN BEHCET'S SYNDROME PATIENTS WITH OCULAR INVOLVEMENT: DOES ACTUALLY THE DISEASE TEND TO GROW DIM OVER TIME?

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Background: Behçet's syndrome (BS) is a multisystemic, chronic relapsing inflammatory disease classified among the vasculitis. Eye involvement represent one of the most serious manifestation of BS and occurs in half of all patients. It seems more frequent and severe among young males and, unluckily, it still represents a significant cause of morbidity.

Objectives: The aims of the study were: to study the relapse rate of BS patients with ocular involvement and to identify factors able to predict long-term outcome

Methods: Among a cohort of 138 patients with a diagnosis of BS according the ISG criteria, 62 patients (37 males and 25 females; mean age at the onset 38 $\pm$ 5 years) with ocular involvement were prospectively studied. The probability of clinical relapse after remission of the first ocular attack was calculated using the Kaplan-Meier method. Predictors of long-term outcome were identified by univariate analysis using the log-rank test and by multivariate analysis using Cox proportional hazards regression models.

Results: The mean time between the first initial symptoms of BS and the onset of eye lesions was 2 $\pm$ 2 years. The number of ocular attacks were the following: 43 posterior uveitis, 27 anterior uveitis, 26 retinal vasculitis, while panuveitis developed in 18 subjects. The cumulative relapse rates at 1 year, 3 years, and 5 years after remission of the first ocular attack were 41%, 31%, and 28%, respectively. On multivariate analysis, a younger age (<30 years) at the onset of ocular involvement, male sex and medical treatments other than biological agents represent independent predictive factors for more early relapses in BS patients with ocular involvement.

Conclusions: The relapse rate seems to be more frequent in the first years of diseases, and probably it could be related to the fact the disease tend to grow dim over time. As literature data suggest, younger age and male sex still represent predictive factors of poor long-term clinical outcome.

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AB0696 CLINICAL CHARACTERISTICS AND OUTCOME OF PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS (GPA) ACCORDING TO ANCA POSITIVITY AND SPECIFICITY

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Background: GPA is a necrotizing granulomatous vasculitis associated with ANCA that usually involves ENT, lung and kidneys. ANCA are mostly directed against proteinase 3 (PR3), although in few cases are directed against myeloperoxidase (MPO) or negative.

Objectives: To analyse the phenotype, clinical course and outcome of patients with GPA according to ANCA positivity and specificity

Methods: multicenter retrospective-longitudinal study that included patients diagnosed with AAV between Jan 1995 and Jan 2016 in 21 Hospitals from Spain (REVAS-Study). We analysed the clinical characteristics, treatment and outcome, depending on the ANCA positivity and specificity. Statistical analysis was performed using SPSSv20

Results: 221 patients with GPA were included: 162 with PR3-ANCA, 36 with MPO-ANCA, and 23 with negative-ANCA. The mean-age at disease onset was higher in patients with MPO-ANCA (59.9 $\pm$ 16.5 years) than in patients with PR3-ANCA (50.5 $\pm$ 16.5) and negative ANCA (48 $\pm$ 14.5), $p=0.006$. Compared to