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.

REFERENCE:

- [1] Mahr A, et al. [Epidemiology and natural history of giant cell arteritis]. *Rev Med Interne*. 2017 Oct;38(10):663–669.

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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.

Disclosure of Interest: None declared

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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

patients with PR3-ANCA, patients with MPO-ANCA presented a lower prevalence of toxic syndrome (42.9% vs. 55.6%, $p=0.021$), arthralgias (34.3% vs. 59.9%, $p=0.008$), arthritis (8.6% vs. 26.5%, $p=0.026$), pulmonary involvement (cavitated/infiltrated nodules, $p=0.007$ and $p=0.05$), and anaemia (57.1% vs. 77.8%, $p=0.05$). Renal disease was less severe in patients with MPO-ANCA (creatinine ≥ 1.58 mg/dL 14.3% vs. 30.4%, $p=0.04$). Subglottic stenosis and sensoryneural deafness were more frequent in patients with MPO-ANCA (20% vs. 6.8%, $p=0.05$ and 34.3% vs. 18%, $p=0.03$). The mean BVAS at baseline was lower in patients with MPO-ANCA (16.3 $\pm$ 8.8) and negative ANCA (12.5 $\pm$ 6.7) than in patients with PR3-ANCA (19.6 $\pm$ 8.9), $p=0.029$. Patients with negative ANCA had less frequently toxic syndrome, fever, arthritis, subglottic stenosis, kidney disease, and peripheral neuropathy, and more frequently orbital masses. Disease relapses were less frequent in patients with MPO-ANCA than in patients with PR3-ANCA (37.1% vs. 48.8%, $p=0.04$), but more frequent than in patients with negative ANCA (20.8%), $p=0.002$. Patients with MPO-ANCA and negative ANCA received less frequently oral cyclophosphamide. No significant differences were found related to death in patients with MPO-ANCA and PR3-ANCA. Patients with negative ANCA had the lower mortality.

Conclusions: A small percentage of patients with GPA present MPO-ANCA or negative ANCA. In our series, patients with MPO-ANCA were older at the disease onset, presented limited or less severe organic disease than patients with PR3-ANCA, lower percentage of relapses and lower requirement of aggressive therapies. Patients with negative ANCA had the best prognosis. Our findings are similar to those recently published,¹ although our patients with MPO-ANCA were older. Classification of GPA patients considering ANCA specificity can improve the treatment stratification and reduce adverse events.

REFERENCE:

- [1] Schirmer JH, et al. Myeloperoxidase-antineutrophil-cytoplasmic-antibody (ANCA) positive Granulomatosis with polyangiitis. A clinically distinct subset of ANCA-associated vasculitis. *Arthritis Rheum* 2016;68:2953–63

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AB0697 GIANT CELL ARTERITIS WITH NORMAL INFLAMMATORY MARKERS AT DIAGNOSIS

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Background: GCA is an inflammatory vasculitis affecting medium and large-sized arteries, that can result in arteritic anterior ischaemic optic neuropathy. C-reactive protein (CRP) level and erythrocyte sedimentation rate (ESR) are usually elevated at GCA diagnosis, but inflammatory-marker negative disease does occur.

Objectives: To analyse the clinical and histological findings of patients with biopsy-proven GCA and negative inflammatory-markers at diagnosis.

Methods: multicenter-longitudinal retrospective study that included patients with biopsy-proven GCA recruited at 10 Hospitals from Spain (REVAS Study). Statistical analysis was performed using SPSS vs. 21.

Results: 418 patients: 290 (69.4%) females (ratio F/M: 2.3/1) were included. The mean age at diagnosis was 75.5 $\pm$ 7 (53–92). The most frequent symptoms at diagnosis were recent onset headache (81%), toxic syndrome (47%) and rheumatic polymyalgia (44.5%). Jaw claudication, cranial hyperesthesia and amaurosis fugax were reported by 44.5%, 31.8% and 16.5% of patients, respectively. A total of 84 patients suffered permanent vision loss. Fourteen patients (3.3%) had normal ESR (<40 mm/h) and CRP (<5 mg/L) at diagnosis. No significant differences were found related to age at disease onset in these patients. Most patients (85%) reported headache; 42.9% jaw claudication, 28.6% cranial hyperesthesia and 42% rheumatic polymyalgia. Fever was less frequent in patients with negative inflammatory-markers (7.7% vs. 40.4%, $p=0.020$), as well as toxic syndrome (21.4% vs. 51.5%, $p=0.031$). In contrast, patients with negative inflammatory-markers had more frequently amaurosis fugax (35.7% vs. 16.8%, $p=0.03$) and optic ischaemic neuropathy (50% vs. 18.7%, $p=0.009$). Temporal arteries were abnormal (thickened and/or pulse less) in 78.6% of patients with negative inflammatory-markers vs. 37% of patients with elevated ESR and/or CRP. Anaemia was less common in patients with negative inflammatory-markers (28.6% vs. 81.5%, $p<0.001$, mean haemoglobin 12.9 $\pm$ 1.1 g/dL). No significant differences were found related to temporal artery biopsy findings, although patients with normal ESR and CRP showed giant cells in 74.3% of cases vs. 62%.

Conclusions: typically patients with GCA present with elevated inflammatory-markers (ESR and CRP) at disease onset. However, a few percentages of patients (4%–5%) have normal ESR and CRP at diagnosis.¹ In our series, 3.3% of patients had negative inflammatory-markers at diagnosis. These patients had fewer constitutional symptoms and more visual symptoms (amaurosis fugax and permanent visual loss) than patients with elevated ESR and/or CRP. Our result are similar to those published in the literature. Abnormal temporal arteries on physical examination may help to diagnosis

REFERENCE:

- [1] Utility of Erythrocyte sedimentation rate and C-reactive protein for the diagnosis of Giant Cell arteritis. *Semin Arthritis Rheum* 2012;41:866–71

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AB0698 A COMPARISON OF THE EFFECTIVENESS OF MYCOPHENOLATE MOFETIL OR METHOTREXATE IN COMBINATION WITH PREDNISOLONE VERSUS PREDNISOLONE ALONE IN THE TREATMENT OF LARGE VESSEL VASCULITIS

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Background: The mainstay of treatment for large vessel vasculitis is glucocorticoids. Immuno-suppressants, including mycophenolate mofetil (MMF) and methotrexate (MTX) are used as steroid-sparing agents.¹ A previous study at our centre showed MMF to have a steroid sparing effect in 97% of patients and to reduce C-reactive protein (CRP) in 80%.²

Objectives: This study was undertaken to compare the efficacy of MMF or MTX combined with prednisolone or prednisolone alone in the treatment of large vessel vasculitis.

Methods: Patients with large vessel vasculitis (LVV) confirmed on positron emission tomography (PET) scan and those meeting ACR criteria for a diagnosis of giant cell arteritis (GCA), treated with prednisolone alone, prednisolone with MMF or prednisolone with MTX started within 3 months of prednisolone being commenced and with a minimum follow up of 24 months were included in a retrospective single centre study. CRP and prednisolone doses were recorded at baseline, after 3, 6, 9, 12, 18 and 24 months of treatment and area under the curve (AOC) calculated for CRP and prednisolone doses. Median AOC prednisolone dose for patients treated with MMF or MTX was then compared with that of patients treated with prednisolone alone. A quantile regression model was also constructed to compare prednisolone dose between the 3 treatment groups, adjusted for CRP. StataCorp. 2013. *Stata Statistical Software: Release 13*. College Station, TX: StataCorp LP was used for all statistical calculations.

Results: 65 patients were included in the study, 41 with GCA and 24 LVV. 49 were female and 16 male. Mean age at diagnosis was 68; range 21 to 87. 37 patients were treated with prednisolone alone; 35 had GCA and 2 LVV. 20 were treated with MMF and prednisolone; 4 with GCA and 16 LVV. 8 were treated with MTX and prednisolone; 2 had GCA and 6 LVV. The AOC for prednisolone and CRP were not normally distributed across the cohort, and non-parametric methods were therefore used for comparisons. Median AOC prednisolone dose for the prednisolone only group was 68.0, (interquartile range (IQR) 17.7, n=37), for the MMF treated group 70.8 (IQR 28.7, n=20) and for the MTX treated group 67.8 (IQR 20.4 n=8). Median AOC CRP was highest in the group treated with prednisolone alone (58.9, IQR 34.5) compared to MMF (43.8, IQR 26.5) and MTX (49.3 IQR 67.5) but there were no statistical differences between median AOC prednisolone dose or CRP in either the unadjusted or regression models.

Conclusions: No significant difference was shown between the groups. MMF is as effective as MTX and prednisolone alone in the treatment of LVV. However, there are limitations to the study. The patient group was small. There was no randomisation to treatment group; treatment choice was based on clinician preference. There was potential bias in that patients perceived to be more difficult to treat may have been given MMF or MTX in addition to prednisolone, and there was a higher proportion of patients with LVV compared to GCA in the MMF and MTX treated groups.

REFERENCES:

- [1] Mukhtyar C, et al. EULAR recommendations for the management of large vessel vasculitis. *Ann Rheum Dis* 2009;68:318–23.
- [2] Smith R et al. Is Mycophenolate Mofetil Effective in the Treatment of Large Vessel Vasculitis? *Rheumatology* 54, Issue suppl_1, 1 April 2015, Pages i194.

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