Methods: Soluble ES and MMP-9 were measured in 68 patients with KD, 20 patients with febrile disease and 20 healthy children by using double antibody sandwich enzyme linked immunosorbent assay (ELISA). Patients with KD were separated into acute phase group, subacute phase group, recovery phase group, coronary artery lesion group (CAL), non-coronary artery lesion group (NCAL).

Results: KS and MMP-9 levels in the acute phase group, subacute phase group were significantly higher than those in the healthy group. Plasma ES and MMP-9 levels of CAL group were significantly higher than those of NCAL group in acute phase. The peak level of ES and MMP-9 appeared in the acute phase. There was a significant correlation between KS and MMP-9 levels in KD patients ($r=0.643$, $P<0.01$).

Conclusions: the increase of plasma PS and ES levels in KD acute phase and subacute phase might play an important role in the pathophysiology of the Vasculux damage. KS and MMP-9 may potentially be a predictor of CAL in patients.

Disclosure of Interest: None declared

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AB0570 CLASSIFICATION, EPIDEMIOLOGY, AND CLINICAL PHENOTYPES OF PRIMARY VASCULITIDES IN COLOMBIA

J.E. Barahona-Correa^{1,2}, Ó.E. Gómez-Cárdenas³, J.C. Sarmiento Monroy^{1,2}, R.D. Mantilla^{1,2}. ¹Rheumatology, Center of Dermatology and Rheumatology (FUNINDERMA); ²Center for Autoimmune Diseases Research (CREA), Universidad del Rosario; ³Department of physiological sciences, School of Medicine, Pontifical Xavierian University, Bogotá, Colombia

Background: Primary vasculitides usually pose a diagnostic challenge. They represent a wide spectrum of heterogeneous vascular disorders characterized by variable target vessel involvement, vascular abnormalities, and end organ damage [1]. Several studies about primary vasculitides expose epidemiologic data, mainly from North America and Europe. Nevertheless, data in Latin America is scarce [2]. **Objectives:** To describe the prevalence and clinical presentation of primary vasculitides.

Methods: This was an observational cross-sectional study in which well-characterized patients were assessed from 2001 to 2016. Each patient was evaluated by the same rheumatologist in a single rheumatology outpatient center in Bogotá, Colombia. Patients were classified either according to ACR 1990 criteria (Granulomatosis with polyangiitis, Eosinophilic granulomatosis with polyangiitis, Henoch-Schönlein purpura, Polyarteritis nodosa, Takayasu arteritis, Giant cell arteritis), EMA algorithm (Microscopic polyangiitis) or 2012 revised Chapel Hill nomenclature criteria (Variable Vessel Vasculitis, Single-Organ Vasculitis). ANCA-Associated Vasculitis (AAV) was used to define Small Vessel Vasculitis (SVV) not fulfilling either ACR 1990 criteria or EMA algorithm.

Results: A total of 56 patients were included. Baseline characteristics of patients were as follows: female gender 75%, mean age 56±15.7 years, and median disease duration 3 (IQR 6) years. Most of patients (73%) were diagnosed

Table 1. Demographic and immunologic characteristics.

Female gender, n [%]	42 [75]
Mean age at onset, years	51 ± 16.7
Mean age at diagnosis, years	53 ± 15.9
Polyautoimmunity*	7 [13]
Multiple Autoimmune Syndrome**	6 [9]
AAV ANCA distribution	n [%]
C-ANCA/PR3*	12 [40]
P-ANCA/MPO	11 [37]
Both ANCA	3 [10]
No data	4 [13]
Type of vasculitis	n [%]
Small Vessel	31 [55]
AAV	19 [34]
Granulomatosis with polyangiitis	8 [14]
Eosinophilic granulomatosis with polyangiitis	2 [4]
Microscopic polyangiitis	1 [2]
Henoch-Schönlein purpura	1 [2]
Medium Vessel	4 [7]
Polyarteritis nodosa	4 [7]
Large Vessel	13 [23]
Takayasu arteritis	5 [9]
Giant cell arteritis	8 [14]
Other	8 [14]
Behçet	3 [5]
Aortitis	1 [2]
Skin-limited	3 [5]
Cogan syndrome	1 [2]

* The presence of more than one autoimmune disease in a single patient. Distribution of autoimmune comorbidities were as follows: antiphospholipid syndrome, systemic lupus erythematosus-SLE like (3 each); autoimmune hepatitis, autoimmune thyroid disease, myasthenia gravis, pyoderma gangrenosum, rheumatoid arthritis, Sjögren's syndrome, and vitiligo (1 each).

** Multiple Autoimmune Syndrome: The presence of three or more well-characterized autoimmune diseases in the same patient.

* A case of TAK with positive C-ANCA was excluded.

at first year of disease onset, and fulfilled international classification criteria (63%). SVV was the most frequent phenotype (55%), and C-ANCA/anti-PR3 were the most frequently identified auto-antibodies (40%). Musculoskeletal manifestations (arthralgia, arthritis, and myalgia), mucocutaneous disorders (including vasculitic/necrotic lesions, mucosal ulcers, and purpura), neurological compromise (peripheral and central), and renal involvement (acute renal failure, glomerulonephritis, and lung-kidney syndrome) were the most frequently reported onset symptoms in 29%, 29%, 23%, and 21%, respectively. Interestingly, most of patients did not develop organic compromise other than the onset manifestation form. Thirteen percent of patients fulfilled polyautoimmunity criteria and 9% presented with multiple autoimmune syndrome. Antiphospholipid syndrome was the most common associated autoimmune disease described. Corticosteroids were the most common treatment used in 93% of patients, followed by azathioprine in 57%, cyclophosphamide, methotrexate, and rituximab in 29%, and antimalarials in 27% (Tab. 1). No deaths occurred during follow-up.

Conclusions: Vasculitides are conditions with several subphenotypes, being ANCA-associated the most frequently reported. Onset symptoms seem to be the main drivers of disease evolution. Appropriate and prompt diagnosis is critical to enable timely intervention, aimed to prevent end organ damage and reduce morbidity in these patients. Controlling disease activity and preventing progression is the milestone of treatment. Characterization of Latin America population is pivotal to raise awareness of health-care workers, and policy makers.

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AB0571 BIRMINGHAM VASCULITIS ACTIVITY SCORE MORE THAN 9.5 AT DIAGNOSIS IS AN INDEPENDENT PREDICTOR OF REFRACTORY DISEASE OF GRANULOMATOSIS WITH POLYANGITIS

J. You, H.J. Kim, S.-W. Lee. Division of Rheumatology, Department of Internal Medicine, Yonsei University College of Medicine, Seoul, Republic of Korea, Seoul, Korea, Republic Of

Background: Granulomatosis with polyangiitis (GPA), that is identical to what has been called Wegener's granulomatosis, is one of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV). GPA is characterized by necrotizing granulomatous inflammation usually affecting small to medium vessels, and it often involves the upper and lower respiratory tracts and commonly provokes necrotizing glomerulonephritis. It has been reported that the mortality rate in untreated patients increased up to 90% within 2 years after diagnosis, and the overall mortality rates were ranging from 12% to 44% within 4 to 10 years. The major causes of death are known as cardiovascular disease, adverse events of immunosuppressive agents and major organ involvement of GPA. If there might be predictors of relapse or refractory disease of GPA during the follow-up duration, they can help physicians to select the induction therapeutic regimens, decide the duration of the maintenance therapeutic regimens and adjust the follow-up interval in order to improve the disease course of GPA.

Objectives: We investigated whether clinical manifestations, anti-neutrophil cytoplasmic antibodies (ANCAs), Birmingham vasculitis activity score (BVAS) for granulomatosis with polyangiitis (GPA) and five factor score (FFS) at diagnosis can predict relapse or refractory disease in 30 histology-proven GPA patients with the follow-up duration ≥ 12 weeks.

Methods: We reviewed the medical records of 30 GPA patients. We collected clinical data, ANCAs, BVAS for GPA, FFSs at diagnosis, and we compared variables between the two groups based on relapse or refractory disease. The optimal cut-offs were extrapolated. Multivariate logistic regression and Cox hazard model analyses were conducted to identify predictors of refractory disease.

Results: The mean age and follow-up duration of patients were 63.3 years old and 45.2 months. The mean initial BVAS for GPA, FFS (1996) and FFS (2009) were 5.4, 0.6 and 1.0. There were no significant predictors of relapse. The mean BVAS for GPA, FFS (1996) and FFS (2009) of patients with refractory disease were higher than those without ($p<0.05$ for all). Patients having BVAS for GPA ≥ 9.5, FFS (1996) ≥ 2 and FFS (2009) ≥ 2 exhibited significantly enhanced risk of refractory disease than those having not (RR 23.0, RR 11.0, and RR 55.0, respectively), and low cumulative refractory disease free survival rates. Multivariate Cox hazard model analysis proved BVAS for GPA ≥ 9.5 was an independent predictor of refractory disease during the follow-up duration (OR 12.892).

Conclusions: BVAS for GPA ≥ 9.5 was an independent predictor of refractory disease during the follow-up duration ≥ 12 weeks.

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AB0572 EFFICACY AND SAFETY OF RITUXIMAB FOR ANCA ASSOCIATED VASCULITIS

K. Fujita¹, M. Hirata¹, S. Oguni², Y. Nakashima², Y. Nomura², Y. Yamamura³, K. Nonaka², N. Shibutou^{1,2}, N. Momoki², A. Ueno¹, T. Ombe², K. Maruyama², M. Hiramatsu², M. Yamamura¹. ¹Centre for Rheumatology; ²Centre for Kidney Diseases, Okayama Saiseikai General Hospital; ³Okayama University Hospital, Okayama, Japan

Background: In Japan, rituximab (RTX) has become one of the dominant alternatives for ANCA associated vasculitis (AAV) even for elderly patients.

Objectives: The efficacy and safety of RTX for AAV patients was determined at our hospital.

Methods: Nineteen patients with AAV, including 14 newly diagnosed patients and 6 relapsed patients, had been treated with RTX. There were 10 males and 9 females. They were all MPO-ANCA positive; 16 were diagnosed with microscopic polyangiitis (MPA), 2 with granulomatosis with polyangiitis (GPA), and 1 with undifferentiated, according to the EMA classification of AAV. The mean age at RTX induction was 71.3 years (range: 40–82 years). The efficacy was evaluated by the BVAS score at the time of first induction and after 6 month treatment. Adverse events were recorded during the 6-month treatment.

Results: The mean of BVAS decreased from 17.3 (range: 7–35) at the first induction to 1.2 (0–4) at 6 month of RTX treatment. Of the 10 patients who could be followed-up for over 6 months, 7 patients achieved remission (BVAS=0) (remission rate: 70.0%). The titer of mean MPO-ANCA decreased 136.1 IU/mL (9.9–300 IU/mL) to 44.4 IU/mL (1.0–114.0 IU/mL) at 6 month. The dose of prednisolone decreased from 34.4mg/day (5–60mg/day) at baseline to 5.5mg/day (0–10 mg/day) at 6 month. The adverse events were as follows: 3 patients experienced reactivation of cytomegalovirus (CMV) with a CMV colitis, one patient with sepsis following urinary-tract infection, and one patient with bacterial pneumonia. One patient with PMA died of exacerbation of the disease itself.

Conclusions: In Japan, AAV, mostly MPO-ANCA-positive PMA, affects elderly persons. The results suggest that RTX may provide a therapeutic option for elderly patients at the age of over 70 years with severe MPA, with an acceptable safety profile and rapid glucocorticoid tapering. However, careful monitoring for infectious diseases will be needed.

Disclosure of Interest: None declared

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AB0573 AUTOLOGOUS STEM CELL TRANSPLANTATION IN THE MANAGEMENT OF REFRACTORY BEHÇET'S SYNDROME: A CASE SERIES FROM A SINGLE CENTRE OF EXCELLENCE

L. Chadwick^{1,2}, D. Thong², J. Nair¹, N. Goodson^{1,2}, R. Moots^{1,2}. ¹Aintree University Hospital; ²Institute of Ageing and CHRONIC Disease, University of Liverpool, Liverpool, United Kingdom

Background: Evidence regarding haematopoietic stem cell transplantation (HSCT) in refractory Behçet's Syndrome (BS) is limited. A systematic review in 2014 identified 20 patients with BS who had undergone HSCT, from which 9 patients underwent HSCT with refractory BS as the primary indication. Of these 20 patients: 15 achieved complete remission, 1 patient achieved partial remission, 2 patients relapsed, 1 patient died and 1 had no outcome reported. Nine patients had reported complications, 5 patients had no complications and 6 patients were missing data. All data was sourced from single case studies (1).

Objectives: Outline the indications, outcomes and complications of HSCT in a case series of 4 patients with BS managed at a single centre of excellence.

Methods: Case notes were reviewed for 4 patients from the Liverpool Behçet's Centre of Excellence (LBCE) who have undergone HSCT.

Results: The primary indication for 3 of the 4 patients who have undergone autologous HSCT at LBCE was refractory BS. Patient 1 underwent HSCT for multiple myeloma, but suffered from severe BS with neurological and venous involvement and is now in partial remission with occasional mild muco-cutaneous ulceration, not requiring systemic immunosuppression. Patient 2 underwent HSCT for severe refractory neuro-Behçet's. This patient is now in complete remission after commencing azathioprine for oral ulceration. The indications for HSCT in patients 3 and 4 were severe muco-cutaneous ulceration and dermatological involvement refractory to numerous biologic agents with high ongoing steroid dependency. Patient 3 also had previous thrombophlebitis and patient 4 had previous gastro-intestinal ulceration. They are both currently in complete remission but follow up time is limited. Patient 3 had a post-HSCT complication of pneumonia and mucositis which resolved without persistent morbidity, patient 4 had an upper limb deep vein thrombosis (DVT) as an inpatient.

Conclusions: HSCT is an effective treatment modality for severe refractory BS and can result in medication free remission. However, the associated risks should be considered and alternative treatment options deliberated or exhausted before opting for this treatment modality.

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AB0574 VENOUS INVOLVEMENT IN BEHCET'S DISEASE: A RETROSPECTIVE ANALYSIS OF 75 CASES

L. Belhadj, A. Hariz, I. Boukhris, I. Kechaou, S. Azzabi, L. Ben Hassine, E. Cherif, N. Khalfallah. *Internal medicine B, Charles Nicolle Hospital, Tunis, Tunisia*

Objectives: Behcet's disease (BD) is a chronic multisystem inflammatory disease characterized by recurrent mouth and genital ulcerations. The vascular involvement varies from 7, 7 to 43% depending on ethnicity. Venous lesions are

Abstract AB0573 – Table 1

Patient	Age at HSCT (years)	Sex	Disease duration at HSCT (years)	BS manifestations	Treatment pre-HSCT	HSCT Indication	Follow up (months)	Complications	Outcome	Treatment post-HSCT
1	47	M	6	OU GU S MSK V N	Tacrolimus	Multiple Myeloma	135	Nil	OU GU (minor)	Pred 4mg
2	29	M	5	OU GU S Ocular N	MMF Tacrolimus IFX ETN CYCP ALN	Neuro-Behçets	80	Nil	1x flare 2013 (OU)	AZA
3	51	M	4	OU GU S V N	AZA MMF ETN ADA Certolizumab	Refractory, severe OU & GU	1	DVT	Remission	nil
4	42	F	Unknown	OU GU S MSK Gastro	Interferon AZA Leflunomide MTX Thalidomide IFX ADA ETN ALN	Refractory, severe OU & GU	8	Pneumonia & Mucositis	Remission	Pred 40mg

OU = oral ulceration, GU = genital ulceration, MSK = musculoskeletal, S = skin, N = neurological, V = vascular. AZA = azathioprine, CYCP = cyclophosphamide, MMF = mycophenolate mofetil, MTX = methotrexate. ADA = adalimumab, IFX = Infliximab, ETN = Etanercept, ALN = Alemtuzumab.