damage indices at presentation. One rare but life-threatening complication of AAV is diffuse alveolar hemorrhage (DAH).

Objectives: To study the clinical features, course and response to therapy in Hispanic American patients with AAV and DAH seen at a large public teaching hospital that serves medically underserved and low income patients with chronic poor health maintenance.

Methods: Among 60 Hispanic American patients with AAV, we retrospectively studied 13 patients with DAH from 2003-2017. Clinical features, laboratory results, diagnostic modalities and treatments given were studied.

Results: A total of 13 patients were identified; 6 men and 7 women, mean age of 51 ± 11 years. Anti-PR3 was found in 5 patients, anti-MPO in 8. Nine patients were diagnosed with microscopic polyangiitis and 4 with granulomatous polyangiitis based on ACR criteria or Chapel Hill Consensus Conference definitions. DAH was a presenting manifestation in 12 patients. Gross hemoptysis with hypoxia, drop in hematocrit and chest X-ray infiltrates were seen in all patients diagnosed with DAH. Bronchoscopy with BAL washings confirming diagnosis was done in 10 patients. All patients had multi-organ involvement including nephritis ($n=8$), peripheral neuropathy ($n=1$), palpable purpura ($n=2$), upper respiratory tract involvement ($n=4$) and eye involvement ($n=2$). Serum C3 was normal in 12 patients. Renal biopsy in 9 patients showed pauci-immune crescentic glomerulonephritis. Mean BVAS at time of DAH was 21.2 ± 5.1 .

All patients underwent induction-remission therapy with pulse methylprednisolone followed by tapering oral prednisone. Induction therapy included IV cyclophosphamide ($n=6$), rituximab ($n=6$) and IV cyclophosphamide/rituximab combination therapy ($n=1$). Nine patients underwent plasma exchange and 5 received IV gamma globulin. Eleven patients required mechanical ventilation; 6 patients needed hemodialysis, 2 temporarily.

All 13 patients responded to induction therapy; mean post-induction therapy BVAS was 9.3 ± 3.5 . Pulmonary fibrosis after DAH was noted in 2 patients. No early deaths (<6 months) were observed, however 1 patient died within 3 years due to infection. Mean follow-up period was 69 ± 54 months. Maintenance therapy used included prednisone ($n=13$), mycophenolate mofetil ($n=2$), azathioprine ($n=6$) and IV rituximab ($n=8$).

Five patients had clinical relapse, 3 with recurrent DAH. Two patients had minor relapse, defined by their treating physician as requiring increased prednisone dose or worsening proteinuria. Mean BVAS at relapse was 15.2 ± 5.5 .

Conclusion: This first in-depth study of DAH in Hispanic American AAV patients shows that in this socioeconomically disadvantaged patient population, the response to induction therapy is not significantly different from other ethnic groups reported in the literature. DAH remains a severe clinical manifestation of AAV and should be recognized without delay. If treated promptly following EULAR/ERA-EDTA recommendations, good short term response is to be expected.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.444

AB0617

CUTANEOUS POLYARTERITIS NODOSA TREATMENT: A RETROSPECTIVE CASE SERIES

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Background: Cutaneous polyarteritis nodosa (CPAN) is a small-to-medium vessel vasculitis limited to the skin. As it is a rare condition, there is little data available to guide treatment. Previous articles have suggested that colchicine and dapsone be used in mild disease and glucocorticoids in acute flares, with DMARDs such as methotrexate or azathioprine being reserved for moderate to severe disease, or steroid-refractory disease.

Objectives: Our objective was to review therapeutic regimens used to treat CPAN and to evaluate patient response.

Methods: A retrospective chart review including records from January 2003 to June 2017 was performed to identify patients with both histologic and clinical features of CPAN. We extracted information regarding patient age, clinical presentation, biopsy results, and response to treatment.

Results: Thirteen patients were identified who had clinical features of CPAN that were supported by histologic findings on biopsy. The majority of patients were female (92.3%). The average age at diagnosis was 44.9 years and mean duration of follow up was 53 months. All patients had involvement of the lower limbs with the most common cutaneous manifestation being subcutaneous nodules (92.3%), followed by papules (46.2%) and livedo reticularis (30.8%). Eight out of thirteen patients (61.5%) received steroids and three did not experience clinical benefit. Another patient did not respond to prednisone and hydroxychloroquine, but had significant improvement of her symptoms on prednisone and methotrexate. Seven out of nine patients (77.8%) who were treated with methotrexate had a favourable response. Three of those patients responded to methotrexate after failing other therapies such as dapsone, azathioprine, and prednisone.

Conclusion: In this case series, we found that most patients had a favorable response to methotrexate and thus the role of methotrexate in treating CPAN merits further investigation. As CPAN often has a chronic relapsing and remitting course, initiating methotrexate at the same time as steroid therapy may lower overall lifetime exposure to steroids.

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Disclosure of Interests: Mohan Stewart: None declared, Ada Lo: None declared, Kamran Shojania: None declared, Sheila Au: None declared, Jan Dutz: None declared, Michael Seidman: None declared, Jon Chan Grant/research support from: Janssen, UCB, Novartis, Pfizer, Celgene, Consultant for: Amgen, Celgene, Eli Lilly, Janssen, Amgen, Abbvie, Novartis, Pfizer, UCB, Sandoz, Merck,

DOI: 10.1136/annrheumdis-2019-eular.6353

AB0618

A SNAPSHOT OF DISEASE ACTIVITY IN PATIENTS WITH BEHCET'S SYNDROME: PREDICTIVE FACTORS AND RELATIONSHIP WITH QUALITY OF LIFE

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Background: Behçet's Syndrome (BS) is globally characterized by a variable spectrum of disease profile: while prevalent muco-cutaneous lesions and arthritis represent the only clinical features in patients with a benign disease subset, there are other patients who develop potentially sight or life-threatening manifestations, due to ocular, neurological or major vascular involvement. Beside the organ involvement, demographic factors could considerably influence the long-term and short-term outcomes of BD

Objectives: The primary aim of the study was to evaluate disease activity in a cohort of BS patients consecutively followed in a BS clinic of a tertiary centre; the secondary aims were to identify potential predictive factors of disease activity and to compare disease activity with quality of life.

Methods: One-hundred and thirty patients (71 males and 59 females; mean age 42 ± 8 years, mean disease duration 11 ± 4) with a diagnosis of BS according to the ISG criteria were studied. Disease activity has been evaluated by BDCAF and quality of life by the Short Form (SF) 36. Patients were also categorized in major or minor involvement of BS according or not to the presence of ocular, neurological and vascular involvement in the course of disease 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: At time of the evaluation, 39% of BS patients presented an active disease. Thirty-six patients presented muco-cutaneous involvement, 21 ocular disease, 10 joint involvement, 6 neurological impairment and 4 gastro-enteric involvement. Sixty-two percent of patients presented in the course of the disease a severe BS involvement and, among these, the majority was represented by males HLAB51 positive patients. The major limitations of quality of life related to disease activity were represented by vitality, physical functioning, bodily pain, general health perception.

Conclusion: Younger men patients affected by BS are more suitable to have a more severe disease, due to an increased frequency both of morbidity and mortality, secondary to ocular, vascular and neurological involvement. The variable prognosis associated to different gender or age, may represent an essential and useful element to tailor the management not only to the type and severity of symptoms, but also to the epidemiological profile of BD patients.

Disclosure of Interests: Rosaria Talarico: None declared, Alice Parma: None declared, Eralda Zera: None declared, Emanuele Calabresi: None declared, Chiara Tani: None declared, Chiara Baldini: None declared, Marta Mosca Paid instructor for: GlaxoSmithKline, Lilly, UCB
DOI: 10.1136/annrheumdis-2019-eular.8184

AB0619 **FAMILY MEMBERS WITH BS MAY NOT CLUSTER IN SIMILAR CLINICAL CHARACTERISTICS**

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Background: Both genetic and environmental factors may have roles in the pathogenesis of Behcet's Syndrome (BS). Likewise, occurrence of different clinical cluster of BS may be also influenced by the combination of these two factors. But we do not know the weight of the factors during the emergence of the disease and clinical clusters.

Objectives: We conducted a study for evaluating the frequency of family history for BS in different clinical BS clusters. Furthermore, we assessed the hypothesis that "BS patients in the same family generally accumulate in the similar clinical cluster."

Methods: Eighty-five patients and their relatives in our BS cohort were participated to the study. We used International Study Group Criteria for Behçet Disease (ISBD) criteria (1) for classifying our index patients and their first and second degree relatives as BS. We sub-classified BS patient to mucocutaneous, vascular, acne-arthritis-enthesitis, eye, neurologic, intestinal and undetermined clusters (2). Firstly, we re-evaluated the family history of our BS patients. Then, the relatives of the patients with already known BS or with having symptoms related with BS were interviewed with phone. All participants have been informed about the study and we have taken consent from them.

Results: Twenty-two (25.9%) of the patients had first or second degree relatives with BS. Furthermore, 27 (31.8%) of our patient were classified to mucocutaneous cluster. In our study, the BS patient with enthesitis-arthritis-acne cluster had highest frequency (44.4%) of family history. However, there was no apparent accumulation of similar findings in index BS cases and their close relatives (Table).

Conclusion: According to result of these studies, there was no finding that supports the hypothesis "BS patients in the same family generally accumulate in the similar clinical cluster". Still genetic may have major role in the emerging of the BS, such as it is accepted as a member of a group of diseases called MHC-I –opathy (3). However, like most of the diseases, genetic background is not enough for the expression of full-blown disease. Briefly, even in the similar genetic background, multiple and separate hits of environmental or non-genetic factors may take role in the pathogenesis.

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Table. Features of the clusters' family history

Mucocutaneous Cluster	n=27
Family history for BS n(%)*	10/27(37.0)
Mucocutaneous	5(50.0)
Eye	3(20.0)
Vascular	2(20.0)
Eye Cluster	n=18
Family history for BS n(%)*	1/18(5.6)
Undetermined	1(100.0)
Enthesis-arthritis-acne Cluster	n=18
Family history for BS n(%)*	8/18(44.4)
Mucocutaneous	3(37.5)
Enthesis-arthritis-acne	3(37.5)
Neuro-Behçet	1(12.5)
Undetermined	1(12.5)
Vascular Cluster	n=12
Family history for BS n(%)*	2/12(16.7)
Mucocutaneous	1(50.0)
Eye	1(50.0)
Neuro-Behçet Cluster	n=3
Family history for BS n(%)*	0/3(0.0)
Intestinal Cluster	n=1
Family history for BS n(%)*	0/1(0.0)
Undetermined Cluster	n=6
Family history for BS n(%)*	1/6(16.7)
Eye	1(100)

BS: Behçet's syndrome
* First or second degree relatives

Acknowledgement: None
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2019-eular.2254

AB0620 **EFFICACY AND SAFETY OF TOCILIZUMAB IN GIANT CELL ARTERITIS: A MONOCENTRIC REAL-LIFE EXPERIENCE**

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Background: Giant cell arteritis (GCA) is a large-vessel vasculitis that generally affects people older than 50 years¹. Glucocorticoids are the mainstay treatment². Tocilizumab (TCZ) has recently been shown in clinical trials to be effective and safe for the treatment of GCA³.

Objectives: To describe our real-life experience with TCZ in a cohort of GCA patients from a single Italian Centre.

Methods: We included GCA patients diagnosed according to the 1990 ACR criteria, followed-up at our Large Vessel Vasculitis Clinic, and treated with TCZ. We retrospectively evaluated demographic features, disease characteristics, reasons for TCZ introduction and discontinuation, concomitant therapies and response to treatment. For each patient we evaluated potential eligibility for GiACTA trial. For statistical comparisons, we used non-parametric tests.

Results: 40 patients (32 women, mean age 71±10 years) were included. Table 1 summarizes clinical features, tools for GCA diagnosis (imaging and/or biopsy) and reasons for TCZ introduction. Mean disease duration before TCZ introduction was 21.5±29.1 months. Only 12 patients (30%) met both GiACTA inclusion and exclusion criteria at TCZ introduction. 31 patients were followed-up for 3 months, 23 for 6 months, 17 for 12 months. Mean prednisone (PDN) dose at TCZ introduction was 21.9±13.2 mg. Mean PDN dose was 8.4±6.5 mg (p<0.001) at month 3, 4.8±5.6 mg (p=0.002) at month 6 and 2.3±2.3 mg (p=0.012) at month 12. PDN was stopped in 9 patients (2 at month 3, 5 at month 6, 2 at month 12) due to optimal disease control with TCZ alone. At TCZ introduction, 7 patients were already on methotrexate (MTX). MTX was subsequently stopped in 2 patients (1 at month 3, 1 at month 6) due to disease control. TCZ was temporarily held-off in 2 patients due to zoster reactivation and in 2 patients due to mild infections. TCZ was definitively stopped after 2 months (when PDN dose was still high) in 2 patients due to severe infections and after 6 months in 1 patient due to diverticulitis. No patient experienced severe neutropenia or liver enzymes elevation. In no case, TCZ was judged inefficient.