Objectives In this study we compared the clinical efficacy of cyclosporin A (Cyc-A) and interferon-alfa (INF-alfa) in our patients with severe posterior uveitis.

Methods All patients were classified and followed by means of fundus fluorescein angiography. At baseline, Cyc-A and INF-alfa were given in a dose of 5 mg/kg/day and 4,5 million U three times a week, respectively. In case of an attack, a prednisolone treatment course was added to those monotherapy regimen. Twenty-one patients were included in the study. Eight and eleven patients were started on Cyc-A and INF-alfa regimens, respectively. Here, the results at the 3- and 6-month duration were presented.

Results Two of 8 patients treated with Cyc-A were stopped to continue due to inefficacy and switched on INF-alfa therapy. In the same group one patient had gingival hyperplasia, which partially responded to dose tapering, however another patient with azotemia and hypertension had to be changed to other treatment group. Except one, all patients on INF-alfa regimen completed the study with adequate regression of inflammatory activity. In that patient depression unresponsive to drugs led to stop INF-alfa at the end of 6 months.

Conclusion Although it is difficult to make a clear conclusion about the comparative efficacy of INF-alfa and Cyc-A due to small population and short follow-up. But it seems that INF-alfa is more tolerable than Cyc-A in the treatment of Behçet's Disease with severe ocular involvement.

OP0042 COLOUR DUPLEX ULTRASONOGRAPHY IN THE DIAGNOSIS OF TEMPORAL ARTERITIS

¹G Neshet, ²D Shemesh, ¹M Mates, ³M Sonnenblick, ²HB Abramowitz. ¹Internal Medicine; ²Vascular Surgery; ³Geriatrics, Shaare-Zedek Medical Center, Jerusalem, Israel

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Background The diagnosis of temporal arteritis (TA) usually requires a temporal artery biopsy. Recently it has been reported that a periluminal dark halo, detected by colour duplex ultrasonography (CDU) of the temporal arteries, is a characteristic sign of TA, and commonly appears in these patients.

Objectives To examine the predictive value of the dark halo sign in diagnosing TA.

Methods During a period of 2 years 69 patients suspected of having TA were examined by CDU of both temporal arteries, using the Acuson SEQUOIA 512 with a 7 MHz linear array transducer. Temporal artery biopsy was performed in 32 of the patients. The diagnosis of TA was made if a patient had a positive biopsy, or met all of the 3 following criteria: 1. ACR classification criteria were fulfilled, 2. there was a prompt clinical response to 40–60 mg/day of prednisone, 3. no other diagnosis related to the symptoms was made during a follow-up of 6 months.

Results Periluminal dark halo was observed in 24 of the 69 patients. TA was diagnosed in 12 of them, giving a positive predictive value (PPV) of only 50%. No halo was detected in 45 cases. Only 2 of them had TA, giving a high negative predictive value (NPV) of 96%. The sensitivity and specificity of the halo sign for diagnosing TA were 86% and 78%, respectively.

Conclusion The PPV of the halo sign in CDU of the temporal arteries is unsatisfactory for diagnosing TA. However, The NPV is very high. Thus a negative halo sign can practically serve to rule out a diagnosis of TA and precludes the need for a biopsy in most instances.

OP0043 A STUDY OF OVERLAPPING INFECTIOUS, AUTOIMMUNE AND HAEMATOLOGICAL ETIOLOGIES IN CRYOGLOBULINEMIA IN A TERTIARY UNIVERSITY HOSPITAL

¹M Ramos-Casals, ¹O Trejo, ¹M Garcia-Carrasco, ²J Yague, ¹G De la Red, ¹S Jimenez, ¹MI Quenard, ¹R Cervera, ¹J Font, ¹M Ingelmo. ¹Systemic Autoimmune Diseases Unit; ²Department of Immunology, Hospital Clinic, Barcelona, Spain

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Background

Objectives To study the main etiological factors associated to cryoglobulinemia in a university hospital, to determine the percentage of patients with no evidence of associated disease (? essential? cryoglobulinemia) and to analyse the amount of overlapping between the different etiologies.

Methods We analysed 443 consecutive patients with cryoglobulinemia, who tested positive for circulating cryoglobulins in our Department of Immunology between 1991 and 1999. Of these 443 patients, 258 (58%) were women and 185 (42%) men (mean age 54 years, range 14 to 91).

Results Infectious diseases were detected in 331 (75%) patients, autoimmune diseases in 94 (24%), haematological disease in 33 (7%) and essential cryoglobulinemia in 49 (11%). Patients with autoimmune diseases showed a cryocrit higher than 5% (26%) more frequently compared with those with haematological processes (24%), HCV infection (16%) or essential cryoglobulinemia (2%) ($p = 0.003$). HCV infection was found in 321 (73%) patients, HBsAg in 15 (3%) and HIV antibodies in 29 (7%). Forty (9%) patients had Sjögren's syndrome (SS), 30 (7%) systemic lupus erythematosus (SLE), 7 (2%) periarteritis nodosa and 6 (2%) progressive systemic sclerosis. Finally, 16 (4%) patients had a NHL, 3 chronic lymphocytic leukaemia, 3 multiple myeloma and 2 Hodgkin lymphoma. When the overlapping of more than one etiological factor was analysed, we found that most patients with HBsAg or HIV infection were also HCV positive. In addition, we observed a strong overlap between HCV and some autoimmune (50% in SS, 67% in PAN) or haematological (50% of NHL) diseases.

Conclusion HCV infection was the main etiological factor identified, being present in 73% of patients with cryoglobulinemia. Of the systemic autoimmune diseases, SS and SLE were those frequently associated to cryoglobulinemia, and NHL was the most frequent haematological process found. Associated disease was not found in 11% of patients (essential cryoglobulinemia). Finally, we report a strong overlap between certain etiological factors (HCV, SS and NHL).

OP0044 EPIDEMIOLOGY OF VASCULITIS IN THREE REGIONS OF EUROPE

¹RA Watts, ²W Koldingsnes, ²H Nossent, ³MA Gonzalez-Gay, ³C Garcia-Porrua, ⁴G Bentham, ⁵SE Lane, ⁵DG Scott. ¹Rheumatology, Ipswich Hospital, Ipswich; ²Rheumatology, University of Tromsø, Tromsø, Norway; ³Rheumatology, Hospital Xeral-Calde, Lugo, Spain; ⁴Environmental Sciences, University of Tromsø; ⁵Rheumatology, Norfolk and Norwich Hospital, Norwich, UK

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Background The epidemiology of systemic vasculitis (SV) has been poorly documented. Development of accepted criteria and definitions by the ACR and Chapel Hill Consensus Conference (CHCC) permits comparison of data from different areas. We

have reported that Wegener's granulomatosis (WG) is more common in Norwich (UK) than Lugo (Spain).¹

Objectives Our aim was to extend our study comparing the epidemiology of WG, microscopic polyangiitis (MPA), polyarteritis nodosa (PAN), Churg Strauss syndrome (CSS) to include a population in the far north of Europe.

Methods Adult patients with a new diagnosis of SV were prospectively identified in Tromsø, North Norway (population 371,000; latitude 70°N), Norwich (population 420,000; latitude 52°N), and Lugo, North West Spain (population 241,000; latitude 43°N) between 1988–98. WG, PAN and CSS were classified using the ACR (1990) criteria and MPA the CHCC definition. 95% confidence intervals (95% CI) were calculated using the Poisson distribution for the observed number of cases.

Results The results are given in the Table 1. There were 56 patients from Tromsø, 86 from Norwich and 41 from Lugo. In all areas and all disease categories the incidence was greater in men than women and peaked aged 65–74 years.

Abstract OP0044 Table 1

	Tromsø/million (95% CI)	Norwich/million (95% CI)	Lugo/million (95% CI)
WG	10.5 (7.6–14.2)	10.6 (7.8–14.0)	4.9 (2.4–8.8)
CSS	0.5 (0.1–1.8)	3.1 (1.7–5.2)	0.9 (0.1–3.2)
MPA	2.7 (1.3–4.8)	8.4 (5.9–11.5)	11.6 (7.6–17.0)
PAN	4.4 (2.6–7.0)	9.7 (7.0–13.0)	6.2 (3.4–10.5)
Total	13.7 (10.3–17.8)	18.9 (15.1–23.4)	18.3 (13.1–24.8)

Incidence of vasculitis in 3 areas of Europe.

Conclusion We conclude that the overall incidence and pattern of vasculitis in terms of age and sex distribution is similar in the three areas studied. MPA is more common and WG less common in Southern Europe, whilst CSS appears to be more common in Norwich. This study points to the importance of environmental and/or genetic factors in the pathogenesis of vasculitis.

REFERENCE

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OP0045 PROGNOSIS OF THE PULMONARY ARTERIAL ANEURYSMS IN BEHCET'S DISEASE: AN UPDATE

V Hamuryudan, T Er, M Tanverdi, I Fresko, S Yurdakul, H Yazici. *Behcet's Disease Research Center, Cerrahpasa Medical Faculty, Istanbul, Turkey*

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Background In a previous study¹ our group has reported a 50% mortality rate among 24 patients with Behcet's disease (BD) having pulmonary arterial aneurysms (PAA). The delay in diagnosing and initiating treatment for this complication could have a role in this outcome.

Objectives To determine whether the outcome of PAA in our centre has changed since our previous report and to assess the possible factors that might influence the prognosis.

Methods Patients diagnosed as having PAA after 1992 (Group 2) were identified retrospectively. Their clinical findings and outcome were compared with those of the patients in the previous report (Group 1).

Results Fifteen additional BD patients (all male) were diagnosed as having PAA. All were treated with cyclophosphamide and prednisolone. Their mean age (32 ± 1.8 SEM years), disease

duration (51.5 ± 6.8 SEM months), pathergy positivity (92%), HLA B5 status (60%) and other clinical manifestations were comparable to those of the patients in Group 1. The interval from the first symptom of PAA until the initiation of the treatment was significantly shorter in Group 2 compared to Group 1 (1.7 ± 0.6 SEM months vs. 6.4 ± 1.5 SEM months; $p = 0.026$). The mean follow up time after the initiation of treatment did not significantly differ between groups (50.9 ± 8.5 SEM months for Group 2 and 45.3 ± 11.4 SEM months for Group 1). There were 2 deaths in Group 2 (13%) and 14 deaths in Group 1 (58%). A Kaplan-Meier analysis revealed a significantly better survival for patients in Group 2 compared to the patients in Group 1 (log rank = 6.8; $p = 0.009$). When only the patients who had received cyclophosphamide (with or without prednisolone) were considered in the analysis (17 patients from Group 1 and all patients from Group 2), the outcome was still better for the patients in Group 2 (log rank = 3.4; $p = 0.065$).

Conclusion Early recognition and most probably cyclophosphamide usage improve the outcome of PAA.

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OP0046 HISTOPATHOLOGIC FINDINGS IN TEMPORAL ARTERY BIOPSIES (TAB) EXAMINED FOR VASCULITIS. RESULTS FROM A HIGH ENDEMIC AREA FOR TEMPORAL ARTERITIS (TA) IN SOUTHERN NORWAY

¹G Haugeberg, ²R Bie, ³P Quist Paulsen. ¹Department of Rheumatology; ²Department of Pathology; ³Department of Internal Medicine, Vest-Agder Central Hospital, Kristiansand, S. Norway

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Background Southern Norway has been shown to have the highest incidence of TA worldwide.¹ Extended histopathological findings from TABs performed in this area have previously not been presented.

Objectives To examine the frequency and pattern of histopathological findings in TABs performed in patients recruited from a high endemic area for TA.

Methods All registered TABs in the biopsy database at the Department of Pathology at Vest-Agder Central Hospital (Southern Norway) from the 5-year period 1992–96 were examined. All biopsies were re-examined by one experienced pathologist. All slides were stained with HE stain. In addition, positive biopsies were stained with Giemsa and Lendrum stain.

Results TABs were performed in 275 patients of which 271 were regarded as representative (180 women and 91 men; mean age 70.9 ± 11.4 yrs). Vasculitis was found in 59 cases (18 men and 41 women; mean age 71.4 ± 8.2 yrs). Arteries with vasculitis had significantly larger diameters than those without inflammation (1.6 ± 0.7 mm vs. 1.2 ± 0.8 mm, $P = 0.001$), whereas no difference of artery length was found (7.5 ± 3.2 mm vs. 7.6 ± 3.5 mm, $P = 0.77$). Extension of the inflammation throughout the vessel wall was as follows: transmural 55.2%, media and adventitia 22.4%, adventitia 19.0% and media 3.4%. Reparative fibrosis was found in 52.5%, thrombosis in 3.4% and fibrinoid necrosis in 3.4% of the TABs. The following frequencies of different inflammatory cell types in the TABs were found: giant cells 37.3%, macrophages 45.8%, lymphocytes 100%, plasma cells