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

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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

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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

37.3%, mast cells 54.3%, neutrophilic granulocytes 88.2% and eosinophilic granulocytes 25.4%.

Conclusion Histopathologic findings consistent with vasculitis were detected in approx. 1 out of 5 TABs examined in our high endemic area of TA. No difference in biopsy length was found between positive and negative TABs, questioning the importance of recommended large biopsies of 20 mm examining TABs for vasculitis. The types of inflammatory cells were found to be in accordance with previous reports.

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OP0047 FDG-PET SCAN INVESTIGATIONS IN VASCULITIS, OTHER THAN GIANT CELL ARTERITIS OR POLYMYALGIA RHEUMATICA

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Background An increased 18-fluoro-deoxyglucose (FDG) uptake has been described in the large vessels of the thorax and legs in patients with giant cell arteritis (GCA) and/or polymyalgia rheumatica (PMR),^{1,2} using positron emission tomography (PET). This reflects increased glucose metabolism in these inflamed vessels.

Objectives We wanted to expand the FDG-PET technology to other patients with biopsy-proven vasculitis. We wondered if vasculitis could also be visualised in patients with other types of large vessel vasculitis, in patients with medium-sized vessel vasculitis and in patients with different forms of small vessel vasculitis.

Methods Patients with biopsy-proven vasculitis, different from GCA/PMR, who were admitted to the General Internal Medicine Department of the University Hospital Gasthuisberg, Leuven, Belgium underwent FDG-PET scan before eventual treatment with steroids was started. One hour before imaging, 6.5 Mbq/kg FDG were injected intravenously.

Results A total of 18 patients were included: 2 patients had large vessel vasculitis (1 Takayasu's arteritis, 1 periaortitis), 3 patients had medium-sized vessel inflammation (2 Churg-Strauss syndrome, 1 polyarteritis nodosa or PAN) and 13 patients had small vessel vasculitis (7 Wegener's granulomatosis, 2 Henoch-Schönlein purpura, 2 cutaneous polyarteritis, 1 relapsing polychondritis and 1 microscopic polyangiitis).

Aortic FDG-uptake was very intense in periaortitis and rather discrete, but persistent under steroid treatment in Takayasu's arteritis. There was no increased vascular FDG-uptake in patients with PAN or Churg-Strauss syndrome.

Increased FDG-uptake in the lungs was seen in Wegener's granulomatosis patients with lung involvement, as well as in the patient with microscopic polyangiitis and lung involvement. Cutaneous involvement of the legs in small vessel vasculitis is also reflected on FDG-PET scan, as well as bowel involvement in Henoch-Schönlein purpura. (Para) nasal involvement in Wegener's granulomatosis also produces a hot focus on FDG-PET scan.

Conclusion Apart from GCA/PMR, FDG-PET scan can also be used to visualise the inflamed thoracic arteries in those other – but much less frequent – large vessel vasculitides as there are

Takayasu's arteritis and periaortitis. In medium-sized vasculitis, FDG-PET scan seems to be of much less value. In small vessel vasculitis, internal organ involvement can be visualised, especially in the lungs. However, FDG-PET scan can not distinguish pulmonary vasculitic involvement from pulmonary infection, since both conditions induce more local FDG metabolism.

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Anti-phospholipid syndrome

AB0102 ANTIPHOSPHOLIPID SYNDROME (APS) IN CHILDREN

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Background

Objectives To evaluate clinical features and management options of persistent antiphospholipid antibodies (aPL) in children.

Methods Retrospective chart review of patients, seen between 1987 and 2000.

Results Six patients with ages at diagnosis ranging from 9 to 13 years are presented. One of them with primary APS and 3 with SLE and asymptomatic aPL. The clinical manifestations are shown in the Table 1. All of the patients included were positive for lupus anticoagulant and all of them except the case 4 had positive tests for aCL and A β ₂GPI.

Abstract AB0102 Table 1 Clinical manifestations and treatment

	Thrombotic events	Prophylactic treatment	Outcome	Follow up (years)	Recurrences
Case 1	DVT+Pulmonary embolism	Warfarin	Resolved	7	No
Case 2	Transient ischaemic brain attack	Aspirin	Resolved	9	No
Case 3	DVT	Aspirin	Resolved	9	No
Case 4	-	Aspirin	Unchanged	9	-
Case 5	-	Aspirin	Unchanged	8	-
Case 6	-	Aspirin	Unchanged	8	-

Conclusion In paediatric patients with aPL, the risk of thrombosis, their recurrences and the management of oral anticoagulation may be different from adults.