Although renal disease was common only a minority required long-term renal replacement therapy.

It is not over-represented among indigenous people.

Introduction.— Previous studies suggest that the incidence of granulomatosis with polyangiitis (Wegener’s; GPA) increases along a south-north gradient in the northern hemisphere with an incidence rate of 8.0/100,000/year reported for the population of northern Norway. In the present study, we estimated the incidence of GPA among persons born in Greenland.

Methods.— Greenlandic patients with severe rheumatic diseases are routinely referred to university hospital departments in Denmark for treatment. The Danish National Hospital Register was established in 1977 and contains information on all admissions to Danish hospitals. In the Register, we identified all patients treated under a diagnosis of GPA during 1977–2011. Those born in Greenland were identified through the Danish Civil Registration System. For each Greenlandic-born patient, the GPA diagnosis was validated by medical files review.

Results.— Three Greenlandic GPA patients were identified. Two developed GPA in Greenland, and one developed GPA while living in Denmark. The mean annual incidence of GPA among Greenlanders living in Greenland was 1.2/million (95% CI 0.1–4.4) with no differences observed between genders. Annual incidences of 0.8/million (95% CI 0.02–4.4) and 2.8/million (95% CI 0.07–15.4) were calculated for the age groups 0–44 and 45–99 years, respectively. The annual incidence of GPA among Greenlanders living in Denmark was estimated to be 2.8/million (95% CI 0.07–15.5).

Conclusion.— The occurrence of GPA is lower in the predominantly Inuit population of Greenland than among Caucasians living in arctic parts of Norway. This observation demonstrates that the risk of GPA varies across ethnic groups populating the northern-most regions of the world.

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Microscopic polyangiitis in the setting of usual interstitial pneumonia

K. Keogh1, M. Baqir1, U. Specks2, G. Auhgenbaugh1, G. Battaglia2, F. Londrino2, A. Tironi2, S. Monti2, G. Gina2
1. Mayo Clinic, Rochester, Mn, USA
2. University of Brescia, Brescia, Italy

Introduction.— Usual interstitial pneumonia (UIP) is a specific radiographic and histopathologic pattern of interstitial lung disease. When UIP occurs in isolation (UIP-IPF), it has a poor prognosis. UIP can occur in association with connective tissue disease and has been reported in vasculitis. The relationship between microscopic polyangiitis (MPA) and UIP remains ill defined.

Methods.— We conducted a collaborative observational cohort study between two tertiary referral centers. Directed search strategies of clinical databases identified patients with concurrent diagnoses of UIP and MPA between 1989 and 2011. Inclusion criteria for UIP were: radiographic findings and/or biopsy evidence of UIP. The MPA diagnosis was based on the Chapel Hill Consensus Conference definition. We describe the clinical features including radiographic findings and survival data in this cohort.

Results.— We identified 68 patients with a diagnosis of both MPA and UIP. MPA was diagnosed first in nine patients, UIP in 14, and in 45, the diagnoses were made concurrently. Mean age was 68 ± 10 years. Sixty-two of 67 were P-ANCA positive. One was C-ANCA positive. Mean BVAS/WG at diagnosis 4.9 ± 3.3. Seventy-eight percent had renal involvement. Twenty-three percent had alveolar hemorrhage confirmed (n = 12) or suspected (n = 3). Forty of 68 received cyclophosphamide. Median survival from diagnosis of UIP in patients who received cyclophosphamide was 138.2 months (92–281) versus 51.0 months (28–86) for those who did not (P = 0.0008). The use of cyclophosphamide was associated with a 0.32 fold lower risk of death (OR 0.32; 95% CI 0.16–0.64).

Discussion.— In this cohort of UIP and MPA, the two diagnoses were established concurrently in most cases, suggesting that UIP changes usually develop first. Lack of specific data on causes of death is a limitation of our study.

Conclusion.— Our survival data suggest that in contrast to UIP-IPF, immunosuppression improves prognosis in patients with UIP in the setting of MPA. Whether UIP is a possible disease manifestation of MPA or an unrelated co-occurrence remains unclear.

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P50
Incidence of granulomatosis with polyangiitis (Wegener’s) among Greenlanders

M. Faurschou, M. Helleberg, N. Obel, B. Baslund
Department of Infectious Diseases and Rheumatology, Copenhagen University Hospital Rigshospitalet, Copenhagen, Denmark

Introduction.— Previous studies suggest that the incidence of granulomatosis with polyangiitis (Wegener’s; GPA) increases along a south-north gradient in the northern hemisphere with an incidence rate of 8.0/100,000/year reported for the population of northern Norway. In the present study, we estimated the incidence of GPA among persons born in Greenland.

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