Doubled prevalence rates of ANCA-associated vasculitides and giant cell arteritis between 1994 and 2006 in northern Germany

Karen Herlyn¹,², Frederike Buckert¹,², Wolfgang L. Gross¹,² and Eva Reinhold-Keller¹,²

Abstract

Objectives. The aim of this study was to investigate the period prevalences of ANCA-associated vasculitides (AAV), including granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), eosinophilic GPA (EGPA)/Churg-Strauss and GCA, in an urban and rural population in northern Germany in 2006 and to compare the data with our previous study performed in 1994.

Methods. We identified all patients with AAV or GCA via questionnaires to all hospital departments, physicians, health insurance providers, pension funds, reference laboratories for autoimmune diseases and death registries in Luebeck (city) and the rural region of Segeberg (population 468,962) between January and December 2006. The type of vasculitis, gender, year of birth, postal code and death were documented and re-evaluated.

Results. One-hundred and fifty patients were identified, indicating a prevalence of 320 per million inhabitants for the complete catchment area (95% CI 285, 355). GCA was more prevalent than AAV: 171 (146, 197) vs 149 (126, 174). GCA and AAV have almost doubled since 1994. GCA increased from 240 (164, 315) to 440 (399, 481) per million in the population ≥50 years of age and AAV increased from 74 to 149 cases per million. GCA and AAV were more prevalent in the urban compared with the rural region.

Conclusion. The prevalence rates of AAV and GCA almost doubled from 1994 to 2006 for this region with a stable population and using an identical study design. Increased awareness has led to an earlier diagnosis of systemic vasculitis and improved activity-adapted treatment mostly based on randomized controlled trials has led to longer survival. Aspects such as environmental factors and exposure to certain substances need further research.

Key words: ANCA-associated vasculitis, giant cell arteritis, epidemiology, prevalence, granulomatosis with polyangiitis.

Introduction

Regarding the incidence of ANCA-associated vasculitis (AAV), and particularly granulomatosis with polyangiitis (GPA), a substantial number of population-based studies have been performed. Data show rather homogeneous and stable incidence rates in Europe, the USA and New Zealand for GPA, with 8–9 cases/million inhabitants/year [1–8]. Previous studies revealed higher incidence rates in northern countries, including the region Schleswig-Holstein, compared with southern Europe or South America [9]. Similar trends have been observed in GCA [10]. GPA apparently is relatively rare in Japan [11]. Inverse associations of GPA and eosinophilic GPA (EGPA; Churg-Strauss) incidence rates and latitude/ultraviolet (UV) radiation have been reported in a recently published study [12]. Data reporting prevalence rates of AAV (e.g. GPA) are heterogeneous and range between 24 cases/million in France up to 160 cases/million in Sweden [13–16].
An earlier population-based study of our group assessed the period prevalence rates of primary systemic vasculitides for the year 1994 in northern Germany in an urban (the city of Luebeck) and a rural region (Segeberg). The data revealed a prevalence rate for GPA of 58/million inhabitants [15]. Investigations from the UK and Norway reported doubled and tripled prevalence rates for GPA in the last two decades, respectively [1, 16]. Thus this study was conducted to examine the same region and calculate the period prevalence for AAV [GPA, EGPA, microscopic polyangiitis (MPA)] and GCA in this stable population in 2006.

**Methods**

**Catchment area, study population, study period**

Based on an identical retrospective study design as performed in 1994, the prevalence rates of AAV (GPA, MPA, EGPA) and GCA were reassessed in the city of Luebeck and the rural region of Segeberg between 1 January and 31 December 2006. Both study areas are located in the northern German federal state of Schleswig-Holstein. The city of Luebeck is not located within the rural area of Segeberg. The distance between Luebeck and Segeberg is ~30 km. The city of Luebeck had 211,213 inhabitants and 257,749 inhabitants resided in the rural region of Segeberg in 2006. For further details, see Table 1 and Fig. 1.

The city of Bad Segeberg, with a population of 15,000, is the largest city in Segeberg, an area dominated by agriculture and forestry. There are no industrial facilities located in the area. In both areas, the city of Luebeck and the rural region of Segeberg, the population has remained relatively stable over the last 20 years (>99% of the population is Caucasian), with low rates of emigration and immigration (<5%; in the population ≥50 years of age, only 2%). Emigration and immigration are not influenced by the diagnosis of vasculitis. However, the percentage of people ≥50 years of age, both male and female, was significantly higher in 2006 compared with 1994 (see Table 1). More physicians are now registered in both regions: 676 in 1994 in both regions compared with 818 physicians in 2006, corresponding to one physician for every 514 inhabitants vs one physician for every 421 inhabitants in 2006 in Luebeck, and in Segeberg there was one physician for every 915 inhabitants in 1994 vs one for 816 inhabitants in 2006 (Table 1). All data are based on the population statistics for 1994 and 2006 obtained from the Department of Vital Statistics of Schleswig-Holstein.

**Sources**

As in 1994, data on the prevalence of AAV and GCA were collected from the following sources: (i) the hospital departments where the authors are employed (Department

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**Table 1** Characteristics of the study region in 1994 and 2006

<table>
<thead>
<tr>
<th></th>
<th>City</th>
<th>Rural region</th>
</tr>
</thead>
<tbody>
<tr>
<td>Population, n^a</td>
<td>217,133</td>
<td>211,213</td>
</tr>
<tr>
<td>Male/female, %</td>
<td>47/53</td>
<td>48/52</td>
</tr>
<tr>
<td>Population ≥50 years of age, n</td>
<td>84,422</td>
<td>85,338</td>
</tr>
<tr>
<td>Percentage of all inhabitants</td>
<td>39*</td>
<td>40*</td>
</tr>
<tr>
<td>Percentage of all men</td>
<td>34*</td>
<td>37*</td>
</tr>
<tr>
<td>Percentage of all women</td>
<td>43*</td>
<td>44*</td>
</tr>
<tr>
<td>Medical university</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Additional hospitals, n</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>Outpatient clinic for rheumatology</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Physicians outside hospitals, n</td>
<td>422</td>
<td>502</td>
</tr>
<tr>
<td>Non-hospital physicians per inhabitants, n</td>
<td>1:514</td>
<td>1:421</td>
</tr>
</tbody>
</table>

^aP < 0.0001, 1994 vs 2006 for Luebeck; **P < 0.0001, 1994 vs 2006 for Segeberg. ^aData for 30 June 1994 and 31 December 2006.
of Rheumatology, University Medical Centre Schleswig-Holstein, Campus Luebeck and the Department of Rheumatology of the Klinikum Bad Bramstedt; (ii) all departments of the University Hospital of Schleswig-Holstein, Campus Luebeck, including their outpatient clinics; (iii) all other hospitals; (iv) all departments of pathology; (v) all physicians in private practice; (vi) pension funds; (vii) health insurance providers; (viii) death registries and (ix) the reference immunology laboratories serving the catchment area. An additional data source was the Vasculitis Register Schleswig-Holstein, which has continuously collected data on incident cases of systemic vasculitides for the whole federal state of Schleswig-Holstein since 1998 [4].

A detailed information package including the questionnaire sheet was sent by mail to all sources up to three times. In cases with no response to all three mailings, the authors contacted the physicians or hospital departments by phone. The packages included an information sheet for patients with an approval for obtaining further medical details.

Sources (ii), (iii) and (v) were asked to report patients treated because of AAV or GCA. The pathologists identified biopsy specimens with a diagnosis of AAV or GCA in 2006. For positive cases, further information was obtained from the referring physicians or hospitals to confirm the diagnosis. Health insurance companies and pension funds documented patients with AAV and GCA who received medical care in 2006. The central death register was screened to identify patients who died during 2006, regardless of the cause of death. The reference immunology lab results for vasculitis were screened for either a positive ANCA test result or a diagnosis of vasculitis on the request form for all those living in the catchment area in 2006, followed by a detailed evaluation of the medical records.

The study form documented date of birth, gender, second letter of the first and last name, postal code in 2006, type of vasculitis and year of diagnosis. Ethnicity was not documented since the northern German population is 99% Caucasian and has not changed significantly since 1994.

Inclusion criteria

Included were patients with AAV and GCA (ICD diagnostic codes: GPA, M31.3; EGPA, M30.3; MPA, M31.7; GCA, M31.5 and M31.6) living in the catchment area during the defined study period, the entire year 2006. The European Medicines Agency (EMEA) algorithm [17], based on the 1992 Chapel Hill Consensus Conference (CHCC) on the nomenclature of vasculitides, and the ACR criteria for the classification of AAV were applied [17–21]. The CHCC definition and ACR criteria were applied to confirm the diagnosis of GCA [18, 20]. Clinical records were available for most cases and were reviewed, along with all other data, by the authors.

Exclusion criteria

Patients with other types of primary systemic vasculitides than those defined above and all types of secondary vasculitides were excluded, as well as patients not residing in the catchment area in 2006.

Ethics approval

Approval for the study was obtained from the ethics committee of the University of Luebeck. The study was also approved by the data protection agency of Schleswig-Holstein.

Statistical analysis

The period prevalence was calculated as the number of cases per 1 million inhabitants in 2006 for each type of vasculitis separately for the urban and rural region. Gender- and age-standardized (for the population <50 years of age and for the population ≥50 years of age) prevalence rates were also calculated. The 95% CIs were calculated on the basis of the Poisson distribution. The results were compared with a previous study assessing the prevalence rates of AAV and GCA in the identical area in 1994 [15]. The calculations were based on the population statistics of 30 June 1994 and 31 December 2006. Since not all sources were independent, a capture-recapture approach was not performed (the Department of Rheumatology Klinikum Bad Bramstedt and the main immunology reference laboratory are associated).

Results

Two hundred and fifty-one patients were reported from the different sources and 101 patients were excluded from the analysis because they lived outside the catchment area (n = 26), were diagnosed with another type of vasculitis than defined in our inclusion criteria (n = 15), were diagnosed later than 2006 (n = 10) or did not meet the inclusion criteria for other reasons (n = 50).

One hundred and fifty patients with AAV or GCA were identified in 2006, 80 of them with GCA (50 patients in Luebeck and 30 patients in Segeberg), 46 with GPA (24 in Luebeck and 22 in Segeberg), 13 with MPA (9 in Luebeck, 4 in Segeberg) and 11 with EGPA (2 in Luebeck, 9 in Segeberg).

All hospital departments, including their outpatient departments, responded. The rate of written responses from physicians outside the hospitals was 269 of 501 (54%) in Luebeck and 149 of 316 (47%) in Segeberg. Additional telephone calls to those who did not respond to the questionnaires led to information from ~62% of all general physicians and internists. Response rates in 1994 were slightly higher, with 58% for urban vs 55% for rural areas. Data from 44 of 150 patients (29.3%) were obtained from more than one source. Nine patients who died in 2006 were identified from the central death register. The response rate of general practitioners was 19% in Luebeck and 31% in Segeberg in 2006 compared with 33% and 43%, respectively, in 1994. The response rate of internists (17% in Luebeck, 14% in Segeberg) was identical in 1994 and 2006.
Patient characteristics

The median age of all patients was 70 years (range 11–101) in 2006, somewhat higher than in 1994 (66 years, range 4–90). The mean age of patients with GPA was 63.5 years (range 11–80), patients with MPA 61 years (range 37–101) and patients with EGPA 55 years (range 32–89). Patients with GCA had a median age of 74 years (range 56–95).

The median disease duration was 43 months (range 2–296) for GPA, 22 months (range 1–78) for MPA, 42 months (range 6–198) for EGPA and 18 months (range 1–222) for GCA. After informed consents were provided, records were reviewed and data on ANCA testing and biopsies were obtained from 90 patients.

Prevalence of GCA and AAV in 2006

Prevalence rates per 1 million inhabitants for both regions were 98 (95% CI 79, 117) for GPA, 28 (95% CI 18, 117) for MPA and 24 (95% CI 14, 35) for EGPA. The prevalence of GCA was 171 (95% CI 146, 197), but 440 (95% CI 399, 481) for the population \( \geq 50 \) years of age (see Table 2).

Prevalence of GCA and AAV in Luebeck and Segeberg

GPA, MPA and GCA were statistically significantly more prevalent in Luebeck than in Segeberg, while the prevalence of EGPA was significantly higher in Segeberg (see Table 3). In the population \( \geq 50 \) years of age, prevalence rates of GCA were almost twice as high in Luebeck as in Segeberg: 586 (95% CI 539, 633) vs 311 (95% CI 276, 346). This considerable statistically significant difference was also observed in women \( \geq 50 \) years of age [93 (95% CI 835, 951) vs 349 (95% CI 313, 375), \( P < 0.00001 \)], while the observed prevalence rate in men for GCA was higher in Segeberg than in Luebeck [244 (95% CI 213, 275 vs 188 (95% CI 161, 215), not significant] (see Table 3).

**Table 2** Prevalence of ANCA-associated vasculitides and GCA in Luebeck and Segeberg in 2006 compared with 1994

<table>
<thead>
<tr>
<th></th>
<th>Luebeck + Segeberg, 2006</th>
<th>Luebeck + Segeberg, 1994</th>
<th>( P )-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>GPA</td>
<td>98 (79, 117)</td>
<td>58 (36, 80)</td>
<td>0.005</td>
</tr>
<tr>
<td>MPA</td>
<td>28 (18, 117)</td>
<td>9 (0, 18)</td>
<td>0.001</td>
</tr>
<tr>
<td>EGPA</td>
<td>24 (14, 35)</td>
<td>7 (0, 14)</td>
<td>0.001</td>
</tr>
<tr>
<td>GCA</td>
<td>171 (146, 197)</td>
<td>87 (59, 114)</td>
<td>0.001</td>
</tr>
<tr>
<td>( \geq 50 ) years of age</td>
<td>440 (399, 481)</td>
<td>240 (164, 315)</td>
<td>0.00001</td>
</tr>
</tbody>
</table>

Values are given as cases per 1 million inhabitants (95% CI). GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis; EGPA: eosinophilic GPA.

Age- and gender-adjusted prevalence of AAV and GCA

Prevalence rates of GPA were \(~10\)-fold higher in the \( \geq 50 \) year group than in the population <50 years of age: 220 (95% CI 191, 249) vs 21 (95% CI 12, 30) (\( P < 0.00001 \)). Remarkable differences were also found for MPA: 50 (95% CI 36, 63) in the population \( \geq 50 \) years of age vs 14 (95% CI 7, 21) in the <50 years population (\( P < 0.00001 \)). In EGPA, only a tendency has been observed in older vs younger patients: 28 (95% CI 18, 38) vs 17 (95% CI 9, 25), respectively.

Comparing all patients with AAV, no gender differences were found in the population <50 years of age. The prevalence rate for men was 55 (95% CI 41, 69) vs 49 (95% CI 36, 62) for women. For AAV in the age group \( \geq 50 \) years the prevalence rate in men was higher than in women: 328 (95% CI 293, 363) vs 271 (95% CI 239, 303) (\( P < 0.00001 \)). According to the classification of GCA patients, those \(<50 \) years of age were not recruited. The prevalence rate in both regions for GCA was 440 (95% CI 399, 481) in the population \( \geq 50 \) years of age and is almost three times higher in female than in male patients: 612 (95% CI 564, 660) vs 219 (95% CI 190, 248).

Prevalence rates of AAV and GCA in 2006 compared with 1994

The prevalence rates of AAV and GCA assessed in this investigation have at least doubled since 1994. The increases in the prevalence rates of GPA, MPA, EGPA and GCA are statistically significant (Table 2).

Discussion

Between 1994 and 2006 prevalence rates of GPA and GCA doubled in northern Germany. The prevalence rate for GPA rose from 58 to 98 per million inhabitants, the rate for GCA rose from 87 to 171 per million inhabitants and for the population \( \geq 50 \) years of age the rate rose from 240 to 440 per million inhabitants. For MPA and EGPA, prevalence rates in 2006 approximately tripled (9 vs 28 and 7 vs 24, respectively). In contrast, the incidence rates for the same region and Schleswig-Holstein remained stable.

**Table 3** Prevalence of ANCA-associated vasculitides and GCA in Luebeck and Segeberg in 2006

<table>
<thead>
<tr>
<th></th>
<th>Luebeck</th>
<th>Segeberg</th>
</tr>
</thead>
<tbody>
<tr>
<td>GPA</td>
<td>114 (93, 135)</td>
<td>85 (67, 103)</td>
</tr>
<tr>
<td>MPA</td>
<td>43 (30, 56)</td>
<td>16 (8, 24)</td>
</tr>
<tr>
<td>EGPA</td>
<td>10 (4, 16)</td>
<td>35 (24, 46)</td>
</tr>
<tr>
<td>GCA</td>
<td>237 (207, 267)</td>
<td>116 (85, 137)</td>
</tr>
<tr>
<td>( \geq 50 ) years of age</td>
<td>586 (539, 633)</td>
<td>311 (276, 346)</td>
</tr>
<tr>
<td>Male ( \geq 50 ) years of age</td>
<td>188 (161, 215)</td>
<td>244 (213, 275)</td>
</tr>
<tr>
<td>Female ( \geq 50 ) years of age</td>
<td>893 (835, 951)</td>
<td>349 (313, 375)</td>
</tr>
</tbody>
</table>

Values are given as cases per 1 million inhabitants (95% CI). GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis; EGPA: eosinophilic GPA.
from 1998 to 2012, with yearly assessed incidence rates for AAV of 12 new cases per million inhabitants [4, 7]. Similar incidence rates for AAV were documented in Luebeck and Segeberg during this period, with 15.9/million/year in Luebeck and 17.2/million/year in Segeberg. The incidence of GCA was higher in Luebeck, with 27.1/million/year, compared with 14.7/million/year in Segeberg ($P = 0.2$). Constant incidence rates of 8/million/year for GPA were documented. Similar results have been reported for a British population of 3.6 million inhabitants between 1990 and 2005, with a stable incidence of 8.4 (95% CI 7.5, 9.4), while prevalence rates doubled from 28.8/million in 1990 to 64.8/million in 2005 [16]. A recently published Australian study also revealed increased prevalence rates of AAV in the Australian Capital Territory and surrounding New South Wales area between 1995–1999 and 2000–2004. The prevalence increased from 64.3 (95% CI 49.3, 81.7) to 95 (95% CI 76.9, 116.1), while stable incidence rates have been documented: 8.8 (95% CI 4.1, 17.1) and 8.4 (95% CI 3.5, 155.8), respectively [22]. Data from Norway revealed tripled prevalence rates for GPA, from 30/million in 1984 to 95/million in 1998, but also documented an increased incidence of GPA, from 5 to 12/million inhabitants for the same period [1]. Because of the earlier time period examined in the Norwegian study, a comparison of their results and ours, as well as the British and Australian studies, is limited.

Enormous progress has been made since the 1980s with the development of ANCA testing, the implementation of classification criteria and considerable advances in the treatment standards, with stage- and activity-adapted therapy regimens, mostly based on randomized controlled trials. Our results and those of the British and Australian studies starting in the early to mid-1990s (1994–2006, 1990–2005 and 1995–2004) and nearly doubled prevalence rates while the incidence has remained stable are possible indicators of a considerably improved outcome with longer survival and decreased mortality in GPA. Three recently published studies have shown an improved long-term outcome with an almost normal life expectancy for some subgroups of patients with GPA (and other AAV) diagnosed in the 1990s or later [23–25].

The systemic vasculitides analysed in this study are significantly more prevalent in the elderly population compared with the population <50 years of age. The median age of all patients was 70 years in 2006 compared with 66 years in our 1994 study. The higher average life expectancy with a shift towards older average age as documented in our study is also likely to account for the almost doubled prevalence rate, particularly in GCA.

Physician response rates were comparable in 1994 and 2006 and therefore could not explain the increased prevalence rates of AAV and GCA. Similar response rates (53%) were observed in the French prevalence study by Mahr et al. [13]. Furthermore, the majority of vasculitis patients, particularly patients with AAV in Schleswig-Holstein and even other federal states of Germany, are referred to the vasculitis centre in Bad Bramstedt or the University Hospital’s Department of Rheumatology. Finally, improved documentation and software systems in health care allow almost complete registration and recruitment of patients in population-based studies. The collection of data, documentation and registration were similar in 1994 and 2006.

During the same time frame as this investigation, prevalence rates were assessed for GPA in France, New Zealand and Sweden, with heterogeneous rates of 24/million (2000), 152/million (1999–2003) and 160/million (2003), respectively, compared with 98/million (2006) in our study, 64.8/million in Great Britain (2005) and 78/million in Australia (2004) (see Table 4). Considerable differences in prevalence rates of GPA in these countries are obvious despite similar incidence rates in Sweden between 1997 and 2009, with 9.8/million, 8/million in Germany and Great Britain and 8.4–8.8/million in Australia. The particularly low prevalence rates in France may result from the relatively young population in the urban region of Paris and genetic factors within a population with a high proportion of non-Caucasians. Mahr et al. [13] found doubled prevalence rates for AAV in the urban population of Paris of European descent compared with the non-Caucasian population. Epidemiological studies from Japan have revealed that GPA was almost not incident in the observation period from 2000 to 2004, with prevalence rates of 2.3/million inhabitants >15 years of age [11]. Genetic factors may play a significant role. Recently published studies of cohorts from Germany and the UK have revealed a three to four times increased association of GPA with the HLA-DPB1 allele. These findings were not confirmed for MPA or EGPA, leading to the assumption that similar AAVs have different genetic risk factors [26].

Limited and heterogeneous data are available for the epidemiology of EGPA and MPA. A French study revealed prevalence rates for EGPA of 10.7/million in 2000 [13], data from Sweden showed 14 (2003) vs 7 (1994) and this study found a rate of 24 (2006) in Germany [14, 15]. Prevalence rates of MPA in Sweden and New Zealand are significantly higher: in 2003, 94 and 58/million, respectively, compared with 28/million in northern Germany in 2006 [14, 27].

Aetiological factors for these significant differences remain unclear; differences in medical care are unlikely. Methodological issues are possible, e.g. application of the different classification criteria or, as in our study, the algorithm for the classification of AAV [17], or differences in the continuous and complete documentation of medical and health care issues (e.g. in Sweden compared with other countries). Different documentation systems are particularly relevant in prevalence studies in contrast to investigations assessing the incidence of usually severe diseases such as vasculitides. Those patients are very likely diagnosed as inpatients because of the severe and often life-threatening courses of their disease. Therefore complete capture of the incident cases by one reporting source is simple.

Little data regarding prevalence rates for GCA have been published, while several studies report incidence
### Table 4 Prevalence of ANCA-associated vasculitis in Germany compared with other countries

<table>
<thead>
<tr>
<th>Reference no.</th>
<th>Study area</th>
<th>Study period</th>
<th>GPA, cases per 1 million inhabitants (95% CI)</th>
<th>MPA, cases per 1 million inhabitants (95% CI)</th>
<th>EGPA, cases per 1 million inhabitants (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>14</td>
<td>Sweden</td>
<td>2003</td>
<td>160 (114, 206)</td>
<td>94 (58, 129)</td>
<td>14 (0, 27)</td>
</tr>
<tr>
<td>5</td>
<td>USA</td>
<td>1986-1990</td>
<td>581,000</td>
<td>n.i.</td>
<td>n.i.</td>
</tr>
<tr>
<td>27</td>
<td>New Zealand</td>
<td>1999-2003</td>
<td>464,000</td>
<td>58 (37, 80)</td>
<td>n.i.</td>
</tr>
<tr>
<td>1</td>
<td>Norway</td>
<td>1984-1998</td>
<td>1.1 million</td>
<td>n.i.</td>
<td>n.i.</td>
</tr>
<tr>
<td>13</td>
<td>France</td>
<td>2000</td>
<td>3.6 million</td>
<td>25.1 (16, 34)</td>
<td>10.7 (5, 17)</td>
</tr>
<tr>
<td>16</td>
<td>United Kingdom</td>
<td>1990-2005</td>
<td>450,000</td>
<td>n.i.</td>
<td>n.i.</td>
</tr>
<tr>
<td>15, this study</td>
<td>Germany</td>
<td>1994-2006</td>
<td>450,000</td>
<td>1994: 9 (0, 18)</td>
<td>1994: 7 (0, 14)</td>
</tr>
</tbody>
</table>

- GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis; EGPA: eosinophilic GPA; n.i.: no information.

*Population ≥ 15 years of age. *b*Only hospitalized cases or deaths (no information about the confidence interval).*
rates. Two studies, from 1975 [28] and 1984 [29], examined the prevalence of GCA in Olmsted County, MN, USA and found remarkably higher prevalence rates of 1330/million in the population >50 years of age on 1 January 1975 and 2230/million inhabitants on 1 January 1984, compared with our results of 440/million in 2006. Corresponding to the prevalence rates in Olmsted County, significantly higher incidence rates than in our region have been reported (186/million vs 25–45/million). Awareness of GCA is probably higher in the region around the Mayo Clinic than in our region [30].

Similar to our previous study, statistically significant differences in the prevalence rates between the city of Luebeck and the rural region of Segeberg were found. While these differences were observed only in GCA in 1994, this study showed significantly higher prevalence rates for GPA, MPA and GCA for all patients and women >50 years of age in Luebeck vs Segeberg; however, for EGPA and for GCA in men >50 years of age an inverse ratio was seen (see Table 3). The causes for these differences remain unclear; demographic factors, with a higher proportion of inhabitants ≥50 years of age in Luebeck vs Segeberg (see Table 1) may play a role, since the prevalence rates for GCA and AAV are considerably higher in the older population. The density of physicians and specialist physicians as well as the university medical centre may lead to greater awareness regarding systemic vasculitides in Luebeck. In contrast to these data, an Australian study revealed higher prevalence rates for GPA (129.7/million) in rural regions compared with urban areas (81.6/million). Environmental factors and exposure to certain substances may play a role [31, 32].

Previous studies suspected associations with occupational exposures in agriculture, e.g. silica dusts or solvents. However, those studies were performed retrospectively in patients with a confirmed diagnosis of vasculitis and therefore are likely to be overestimated because of recall bias. Knight et al. [32] recently assessed environmental factors and occupational exposures in patients with GPA and did not identify significant risk factors.

In summary, this study observed GCA as the most prevalent systemic vasculitis in 2006 in a northern German population, with 171/million inhabitants (440/million in the population ≥50 years of age), followed by GPA with a prevalence rate of 98/million. Compared with our previous study of the identical population in 1994, the prevalence rates of AAV and GCA have at least doubled while the incidence remained stable. In conclusion, these results show an improving prognosis for systemic vasculitides.

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