ORIGINAL ARTICLE

Stable Incidence of Systemic Vasculitides in Schleswig-Holstein, Germany

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SUMMARY

Introduction: The authors present data on the incidence of primary systemic vasculitides (PSV) in the northern German state of Schleswig-Holstein from 1998 to 2005.

Methods: Population-based study of all new cases of PSV from 1 January 1998 onward in a region with a population of 2.83 million. The sources of patient data were all hospital departments in the catchment area, including outpatient clinics; all departments of pathology; and the reference immunological laboratories serving the catchment area.

Results: Over this eight-year period, 982 patients with newly diagnosed PSV were identified in Schleswig-Holstein, of whom 273 were diagnosed with ANCA (i.e., anti-neutrophil cytoplasmic antibody)-associated vasculitis (AAV). The incidence of all types of PSV combined was between 38 and 54 cases per million population per year. The incidence of AAV (which includes Wegener's granulomatosis [WG], microscopic polyangiitis [MPA], and Churg-Strauss syndrome [CSS]) was between 9.5 and 16 cases per million per year. WG consistently accounted for two-thirds to three-quarters of all new cases of AAV diagnosed each year.

Discussion: This population-based vasculitis registry designed to capture all new cases of PSV in an eight-year period in a northern German region with 2.83 million inhabitants revealed stable incidence figures for all types of PSV and for AAV. Compared to figures obtained in other studies from small regions or referral centers, the incidence rate of WG (as an illustrative type of AAV) in this study was the same as those in Norway and Sweden, lower than that in the United Kingdom, but higher than those in Spain and in Vilnius (Lithuania). It is unclear whether these differences truly reflect a north-south gradient within Europe or are merely due to methodological differences.

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P primary systemic vasculitides (PSV) are very rare diseases whose epidemiological characteristics have not yet been extensively studied, with the exception of arteritis (1–5). Most vasculitides have risen in incidence in recent years, particularly giant cell arteritis (GCA) (6).

A number of groups have found an increased incidence of Wegener's granulomatosis (WG), microscopic polyangiitis (MPA), and Churg-Strauss syndrome (CSS) since the mid-1980s. This finding was associated with the discovery of the anti-neutrophilic cytoplasmic antibody (ANCA) and the recognition of its diagnostic importance (7, 8). Thus, a retrospective study in Sweden revealed a significant increase in the incidence of WG from 0.77 new cases per million inhabitants per year in 1975 to 11.9 in 2001 (9). Statistics from the United Kingdom and Spain, however, for the time periods 1988 to 1998 and 1988 to 2001, respectively, have shown practically unchanged incidences for ANCA-associated vasculitides (AAV) (1, 4, 10).

The case registries that have existed until now could often be faulted for being limited to a small, and thus perhaps unrepresentative, target region with a small population, or for referral bias due to monocentric registration (1–5). This situation motivated the creation of a vasculitis registry in northern and southern Germany, from 1998 onward, within the framework of the BMBF-sponsored joint project "Vasculitides: Their Etiology, Pathogenesis, Treatment, and Epidemiology."

All newly diagnosed primary systemic vasculitides, as defined by the Chapel Hill Consensus Conference, were registered from January 1, 1998, onward in a population-based study with nearly five million persons inhabiting the catchment area (11). The first two years' results showed no difference between northern and southern Germany in the incidence either of primary systemic vasculitides overall, or of any of the individual diseases in this class (12). As two years was too short a time to collect reliable, population-based epidemiological data on rare diseases, the vasculitis registry was kept in operation in the northern German federal state of Schleswig-Holstein (population 2.83 million). An initial report was published with data from the first five years (13). The goal of the present study is to analyze the results from a total of eight years, from 1998 to 2005, with special emphasis on ANCA-associated vasculitides. The Schleswig-Holstein Vasculitis Registry for the first time enables the reliable, prospective determination of the incidence of PSV in

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TABLE 1

Study area and study population of the Northern German Vasculitis Registry (federal state of Schleswig-Holstein)

	1. 1. 2000	1. 1. 2003
Population (n)	2 766 057	2 823 171
Male/female (%)	49/51	49/51
Age 50 years or older	37%	38%
% of all men	34	35
% of all women	40	40
Medical University (n^{*1})	2	
Hospital departments (n^{*1}) (= 100%)	122	
Internal medicine (n ^{*2})	62 (5%)	
ENT (n)	14 (11%)	
Ophthalmology (n)	2 (2%)	
Dermatology (n)	2 (2%)	
Pediatrics (n)	10 (8%)	
Orthopedics/Surgery (n)	18 (15%)	
Gynecology (n)	3 (2%)	
Neurology (n)	6 (5%)	
Pathology (n)	5 (5%)	

*1 Including outpatient clinics

*2 Including all subspecialties (e.g., nephrology, pulmonology, rheumatology)

Germany and is the largest population-based study to date on the epidemiology of vasculitides.

Methods

Study area and study population

The study area is the state of Schleswig-Holstein, which has 2.83 million inhabitants (*table 1*). The calculated incidences are based on the population figures of January 1, 2000, and on the updated figures of January 1, 2003 (Statistical Office of the state of Schleswig-Holstein). There was relatively little population change attributable to immigration and emigration, the rates of which were 6% and 7%, respectively; for persons aged 50 and above, these rates were even lower (2% and 3%).

Study period

All newly diagnosed cases of PSV in Schleswig-Holstein from January 1, 1998, to December 31, 2005, were entered into the registry.

Sources

Before the Vasculitis Registry was begun, its creation was announced in regional medical publications and official university bulletins as well as by personal letter to all potential participants. All of the relevant institutions agreed to participate in the registry and designated a contact person within the institution. Data on PSV cases were regularly reported by, or requested from, the following sources:

- all hospital departments, including outpatient clinics,
- all pathology departments,
- and the regional immunological reference laboratory.

All "sources" were contacted by letter every three months (up to three times) and asked to report all newly diagnosed cases in the preceding three months. If all three letters went unanswered, the institutional contact person for the registry was contacted by telephone. All reported cases were re-evaluated by the authors with regard to the diagnosis, site of residence of the patient, and other features.

As required by data protection guidelines, the initial reporting of each case was anonymous but included the postal code of the patient's residence as well as the patient's sex, date of birth, and diagnosis, and the second letters of the patient's first and last names. If the patient consented, further medical data were obtained, e.g., the nature and manner of onset of the initial symptoms, the organs that were affected at the time of diagnosis, the ANCA findings, the histological findings, and the treatment provided.

All newly diagnosed patients were provided with information about the disease and its treatment and complications, as well as suitable literature for patients, patient education sessions, and contact persons in the nationwide Vasculitis Self-Help Group, which has been in existence in Germany since 1994. The study protocol was examined and approved both by the Ethics Committee of the University of Lübeck and by the State Data Protection Authority of Schleswig-Holstein.

Inclusion criteria

The following types of PSV, as defined in the Chapel Hill Consensus Conference (CHCC) of 1992 (11), were included in the registry:

- giant cell arteritis (GCA),
- Takayasu's arteritis (TA),
- Wegener's granulomatosis (WG),
- microscopic polyangiitis (MPA),
- Churg-Strauss syndrome (CSS),
- Henoch-Schönlein purpura (HSP),
- (isolated) cutaneous leukocytoclastic angiitis (CLA),
- (classic) panarteritis nodosa (PAN),
- Kawasaki syndrome.

Cases in which primary vasculitis was definitely present – as confirmed, e.g., by histopathological examination – but could not be assigned any of the above diagnoses were entered into the registry as "unclassifiable vasculitis" (UV). This category included, for example, isolated primary CNS vasculitis.

The diagnosis was required to have been made during the period of the study, and the patient was required to have resided within the study area at this point in time.

Exclusion criteria

Patients were excluded from the registry if they had secondary vasculitis (e.g., in the setting of inflammatory rheumatological diseases), vasculitis associated

TABLE 2

Characteristics of all PSV patients and AAV patients (classified by age and sex) in Schleswig-Holstein from 1998 to 2005

PSV type; n (m/f) Age, years (range)*	1998	1999	2000	2001	2002	2003	2004	2005
All PSV	152 (67/85)	138 (56/77)	126 (48/65)	111 (54/57)	115 (51/61)	112 (48/64)	107 (34/64)	121 (51/69)
	62 (4/96)	61 (3/96)	62 (2/90)	62 (4/93)	63 (2/82)	66 (5/89)	63 (3/90)	64 (4/88)
WG	21 (9/12)	18 (11/7)	24 (14/10)	24 (12/12)	33 (18/14)	22 (12/10)	24 (13/11)	25 (15/10)
	61 (25–79)	61 (23–87)	58 (24–85)	58 (22–77)	56 (12–79)	65 (26/81)	55 (20/80)	58 (30/83)
MPA	8 (5/3)	8 (4/4)	9 (6/3)	5 (0/5)	7 (3/4)	5 (2/3)	8 (4/4)	4 (1/3)
	67 (57–80)	66 (55–79)	73 (48–81)	74 (67–76)	63 (36–77)	66 (25/80)	69 (63/78)	74 (39/75)
CSS	0	2 (0/2)	4 (2/2)	4 (3/1)	5 (3/1)	2 (1/1)	6 (2/4)	5 (3/2)
		60 (59–62)	52 (36–68)	57 (46–76)	58 (29–60)	43 (26/81)	50 (26/66)	63 (58/88)

* Median age at time of diagnosis (range).

Unknown sex: 1999, 5 patients; 2000, 13 patients; 2002, 3 patients.

PSV=systemic vasculitides; WG=Wegener's granulomatosis; MPA=microscopic polyangiitis; CSS=Churg-Strauss syndrome. m=male, f=female.

with an infectious disease (including hepatitis-associated vasculitis), or vasculitis associated with malignancy.

Statistics

The incidence figures were calculated as the number of new cases per million persons per year, and 95% confidence intervals were determined. The latter intervals were obtained by assuming that the incidence of rare diseases (both PSV as a whole and the individual entities that it comprises) follows a Poisson distribution. Furthermore, stratified incidence figures were calculated by age and sex, with separate calculations for the population subgroups above and below age 50. The number of sources reporting each individual case was recorded. For cases of AAV, it was also studied how frequently ANCA testing was performed in the Bad Bramstedt immunological reference laboratory, and what percentages of the patients were treated as either in- or outpatients in the Bad Bramstedt/Lübeck Vasculitis Center.

Results

A total of 1260 cases were reported from 1998 to 2005. 278 cases were excluded from the registry because the diagnosis was made before 1998, because the patient did not reside in the study area, or because the diagnosis did not correspond to the inclusion criteria. Thus, the registry finally included 982 patients (tables 2 and 3 and e-figure). The numbers of cases per year were as follows: in 1998, 152 cases; in 1999, 138 cases; in 2000, 126 cases; in 2001, 111 cases; in 2002, 115 cases; in 2003, 112 cases; in 2004, 107 cases; and in 2005, 121 cases.

Patients were reported by an average of two out of three possible sources (range, 1 to 3). For 406 patients (41%), the following laboratory tests were performed in the German reference laboratory for ANCA diagnostic studies in Bad Bramstedt:

- ANCA autoantibodies (indirect immunofluorescence plus ELISA for proteinase 3 and myeloperoxidase);

- antinuclear antibodies;
- double-stranded DNA antibodies (dsDNA-Ab);
- extractable nuclear antibodies (ENA).

ANCA-associated vasculitides

Over the entire period of the study, 273 patients developed ANCA-associated vasculitides (AAV) (i.e., WG, MPA, and CSS), corresponding to 28% of all vasculitides, with the number of cases per year ranging from 28 (in 1999) to 45 (in 2002) (table 2). In the course of these eight years, 191 patients developed WG, consistently accounting for two-thirds or more of all cases of AAV. The annual figures were as follows: in 1998, 21/29 (72%); in 1999, 18/28 (64%); in 2000, 24/37 (65%); in 2001, 24/33 (73%); in 2002, 33/45 (73%); in 2003, 22/29 (76%); in 2004, 24/38 (63%); in 2005, 25/34 (73%). The distribution of AAV patients by age and sex is shown in table 4.

TABLE 3

Schleswig-Holstein Vasculitis Registry 1998–2005 (n = 982)

Vasculitis	n (%)
Wegener's granulomatosis	191 (19.5)
Microscopic polyangiitis	54 (5.5)
Churg-Strauss syndrome	28 (3)
Giant cell arteritis	272 (27.7)
Cutaneous leukocytoclastic angiitis	133 (13.5)
Henoch-Schönlein purpura	117 (11.9)
Unclassified vasculitis	135 (13.7)
Classical panarteritis nodosa	19 (1.9)
Takayasu's arteritis	9 (0.9)
Kawasaki syndrome	12 (1.2)

TABLE 4

Age- and sex-adjusted incidences (with 95% confidence intervals) for all types of PSV and ANCA-associated vasculitides (AAV) in Schleswig-Holstein

	1998	1999	2000	2001	2002	2003	2004	2005
All PSV, age < 50	20.3 (11; 29)	24.8 (15; 35)	24.5 (12; 37)	20.4 (14; 27)	12.4 (6; 19)	15 (5; 25)	18.8 (10; 28)	16 (6; 26)
All PSV, age ≥ 50	114.3 (93; 135)	87.0 (69; 105)	78.4 (61; 96)	74.0 (57; 91)	90 (71; 109)	79 (61; 96)	69.4 (53; 85)	73 (56; 90)
All PSV, male	48.9 (35; 63)	42.2 (30; 55)	42.0 (27; 57)	39.7 (30; 40)	37 (25; 49)	35 (21; 49)	31.2 (21; 41)	32 (22; 42)
All PSV, female	59.4 (44; 74)	53.7 (39; 68)	45.1 (29; 61)	39.3 (29; 50)	42 (29; 55)	42 (27; 57)	44.3 (29; 59)	44 (29; 59)
All AAV, age < 50	3 (0; 6)	0.5	5 (2; 8)	5 (2; 8)	6 (1; 11)	6 (1; 11)	7 (2; 12)	5 (2; 8)
All AAV, age ≥ 50	21 (10; 32)	24 (12; 37)	25 (14; 36)	23 (13; 33)	25 (14; 36)	17 (8; 24)	22 (10; 32)	23 (13; 33)
All AAV, male	10.2 (4; 16)	11 (5; 18)	16 (6; 26)	11 (5; 18)	17.4 (9; 24)	11 (5; 18)	14 (8; 20)	14 (8; 20)
All AAV, female	10.4 (4; 16)	9 (3; 15)	10.4 (4; 16)	12.5 (5; 19)	13 (6; 20)	10 (4; 16)	13 (6; 20)	10.4 (4; 16)

PSV = systemic vasculitides

136 (50%) of the 273 patients with AAV underwent ANCA testing at the Bad Bramstedt reference laboratory. 143 of them (53%) underwent in- or outpatient treatment at the Bad Bramstedt/Lübeck Vasculitis Center. The regional distribution of all 273 patients with AAV over the entire temporal course of the study is shown in *table 3* and in the *e-figure*.

Incidences in northern Germany

The overall incidence of PSV of all types in Schleswig-Holstein varied from 38 to 54 new cases per million inhabitants per year over the course of the study. The annual incidence of AAV from 1998 to 2005 ranged from 9.5 to 16 new cases per million inhabitants per year, with a median figure of 12. WG was consistently the most common subtype of AAV, with an incidence ranging from 6 to 12 new cases per million inhabitants per year (median, 8.5). MPA was

more common than CSS, the least common type of AAV (*table 5*).

Age-related incidences

Both PSV overall and AAV were two to five times more common among persons over 50 years of age than in persons under 50 years of age (*table 4*). The incidence of these conditions rose with age to the same extent in men and women. The mean age at diagnosis for all types of PSV was over 60 years (range, 61 to 66); for WG, 55 to 65 years; for MPA, 63 to 74 years; and for CSS, 43 to 63 years (*table 2*).

Sex-related incidences

The overall incidence of PSV was nearly the same among men and women (slightly higher among women, but the difference was statistically not significant). AAV were more common in men, again without statistical significance) (*table 4*).

TABLE 5

Incidences (with 95% confidence intervals) for primary systemic vasculitides (PSV) in Schleswig-Holstein from 1998 to 2005 (new cases per million inhabitants per year)

	1998	1999	2000	2001	2002	2003	2004	2005
All PSV	54 (39; 68)	48 (34; 61)	45 (34; 61)	40 (31; 50)	42 (31; 52)	39 (19; 58)	38 (25; 51)	38 (25; 51)
ANCA-associated vasculitides								
Overall	11 (5; 18)	9.5 (4; 16)	12 (5; 19)	12 (5; 19)	16 (8; 24)	10 (4; 16)	13.5 (7; 20)	12 (5; 19)
WG	8 (2; 14)	6 (1; 11)	8 (2; 13)	9 (3; 15)	12 (5; 19)	7 (4; 11)	9 (3; 15)	8.5 (3; 15)
MPA	3 (0; 6)	2.5 (0; 6)	3 (0; 6)	2 (0; 4)	3 (0; 6)	2 (0; 4)	3 (0; 6)	1 (0; 3)
CSS	0 (0)	1 (0; 3)	1.5 (0; 3)	1 (0; 3)	2 (0; 4)	1 (0; 3)	2 (0; 4)	1 (0; 3)

PSV=systemic vasculitides; WG=Wegener's granulomatosis; MPA=microscopic polyangiitis; CSS=Churg-Strauss syndrome

TABLE 6

Incidences of ANCA-associated PSV in Europe

	United Kingdom* ¹ (4)	Spain* ² (1)	Norway (9)	Sweden (10)	Lithuania (5)	Germany
Study period	1988–1998	1988–2001	1984–1998	1975–2001	1990–1999	1998–2005
Study area	NHA, Norfolk	Lugo reference center	3 northern districts	Sweden	Vilnius	Schleswig-Holstein
Study type	Hospital	Hospital	Population	Population	Hospital	Population
Study population (n)	413 000	208 271	464 000	8 000 000	470 000	2 830 000
Types of vasculitis studied	WG, CSS, MPA, PAN	WG, CSS, MPA, PAN	WG	WG	WG, CSS	All PSV
Incidences of ANCA-associated vasculitides*³						
Overall	22	12.1	NA	NA	NA	12
WG	10.6	2.9	8.0	7.8	2.1	8.5
CSS	3	1.3	NA	NA	1.3	1
MPA	8.4	7.91	NA	NA	NA	NA

*¹ among persons over age 15 years; *² among persons over age 20 years; *³ new cases per million persons per year; NHA=Norwich Health Authority; NA=data not available; WG=Wegener's granulomatosis; CSS=Churg-Strauss syndrome; MPA=microscopic polyangiitis

Discussion

The Schleswig-Holstein Vasculitis Registry for the first time enabled the collection of reliable data on the incidence of PSV in Germany. With a target population of 2.83 million people and an observation period of eight years, it is the largest prospective population-based study performed to date on the epidemiology of vasculitides.

The overall incidence of PSV was found to vary between 38 and 54 new cases per year per million people. It is thus comparable to the incidence of systemic lupus erythematosus (14) and nearly twice as high as that of systemic sclerosis (15) or multiple sclerosis (16). The ANCA-associated vasculitides (WG, MPA, and CSS) occur in Schleswig-Holstein with an incidence ranging from 9.5 to 16 new cases per year per million people (median, 12). WG accounts for up to 75% of this group of patients; among the remaining 25%, MPA is twice as common as CSS.

Data from other European countries are available regarding the epidemiology of AAV (1–5, 9, 10). There are, however, major methodological differences between the individual studies. The data were often obtained from so-called tertiary centers or in a population-based manner from very small, and therefore unrepresentative, areas (e.g., under-representation of either larger cities or rural areas). Although many of these studies were carried out for long periods of time, some of them recruited a much smaller number of patients; thus, for example, a Norwegian population-based study of the incidence of WG identified only 55 patients with the disease during a period of 15 years. Studies from the United Kingdom and Spain that were carried out for 11 and 15 years, respectively, recruited only 40 (U.K.) and 12 (Spain) patients over the entire period of the study (1, 4, 10). In contrast, there were

191 new cases of WG in Schleswig-Holstein during the eight years of the present study.

In principle, epidemiological studies can be carried out in one of two ways:

- Observation of a small region over a long period of time, with the advantage of a high capture rate
- Observation of a large region over a shorter period of time, with the advantage of a large number of patients in a relatively short time, but with the disadvantage of a potential failure to capture all cases of the disease.

The differences between these two strategies might play a role in the interpretation of a number of the reported findings on vasculitides. In the past 15 to 20 years, there has been increased interest in AAV, and the diagnostic algorithm has changed (17). The most important contributing factors were the introduction of ANCA testing in the late 1980s (18) and the development of classifying criteria and definitions for PSV in the early 1990s. The latter also permit the identification of forms with a relatively benign course that previously would often have escaped detection (19). Thus, comparing the results of this study with those of previous European studies is likely to be difficult.

The authors' study revealed incidence rates of AAV ranging from 9.5 to 16 new cases per million persons per year (median, 12), compared to 22 in the United Kingdom and 12.1 in Spain (1, 4). Classification criteria have a major influence on epidemiological data regarding PSV, particularly as concerns MPA and PAN. In our study, and in the Spanish study as well, the CHCC definitions (11) were used for PSV, because these take immunological and immunohistochemical criteria into account, unlike the classification system of the American College of Rheumatology (ACR) (19). Furthermore, there are as

yet no ACR criteria for MPA. The CHCC definitions for the first time enable a clear-cut distinction between MPA and PAN, two entities with presumably very different pathogenetic features.

WG was the most common type of AAV in this study and in the British study, yet, in Spain, the incidence of MPA was more than twice as high as that of WG (1, 4). In our population-based study with a catchment area containing 2.83 million inhabitants, WG consistently accounted for two-thirds to three-quarters of all cases of AAV. The data in the Spanish study, in contrast, were derived from a tertiary center in a region containing only 238 000 people. The importance of methodological differences in epidemiological studies of AAV is also demonstrated by a study from Sweden on the incidence of small-vessel vasculitides (3). This study by Tidman et al., carried out in a nephrology department, also identified far more patients with MPA (n = 70) than with WG (n = 19).

We found an incidence of WG ranging from 6 to 12 new cases per million inhabitants per year, with a median of 8.5 new cases (table 5). The incidence of WG was thus significantly higher in Schleswig-Holstein than in Spain or Lithuania (2.95 and 2.1 cases per million inhabitants per year, respectively), comparable to the incidence in Norway (8.0) and Sweden (7.8), and somewhat lower than the incidence in the United Kingdom (1, 4, 5, 9, 10) (table 6). These differences (with the exception of the figure from Lithuania) might reflect a north-south gradient across Europe, similar to the one that has been described for AT, unless they are entirely due to methodological differences. Preliminary data from an initial study in the Southern Hemisphere regarding the incidence of WG and MPA in Lima, Peru (population 930 306) from 1990 to 2004 surprisingly show a nearly tenfold higher incidence of MPA (4.1 per million persons per year) than of WG (0.5 per million persons per year).

The demographic data on the population of our catchment area and of our study patients (sex distribution, age at diagnosis) remained fairly constant over the period of the study. It is worth noting that the median age at the time of diagnosis of WG remained roughly 60 years over the entire study period (range of ages at diagnosis: 22 to 87), while the median age at diagnosis was much younger (between 40 and 50) in large WG cohorts that were recruited about 20 years earlier (21–24), even though the time lag from the initial manifestations to the establishment of the diagnosis has become shorter over the years. In the present study, the median time to diagnosis was 3 months (range, 0 to 48 months), while the comparable figures in earlier WG cohorts were 9 months (range, 0 to 288 months) (23) and 15 months (22). This could be due either to a real increase of the age at diagnosis or to increased attention to small-vessel vasculitis in older persons, or both. A limitation of this study might be a less than total capture of patients that were treated entirely on an outpatient basis (and not in the outpatient clinics of hospital departments) and whose diagnoses were established without any histological

examinations or auto-antibody determinations. One can safely assume, however, that such patients are rare among persons with AAV.

Overview

This prospective, population-based vasculitis registry that was kept for eight years in Schleswig-Holstein, an area with 2.83 million inhabitants, revealed an annual incidence of PSV varying from 38 to 54 new cases per million persons per year. ANCA-associated vasculitides had an incidence ranging from 9.5 to 16 new cases per million persons per year and thus accounted for 28% of all vasculitides; their incidence was stable over the eight years of the study. It currently remains unclear whether there are any regional differences in the incidence of WG. The differences that were found might reflect either a true north-south gradient in Europe, perhaps due to different exogenous exposures, or they might merely reflect methodological differences between studies.

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Conflict of interest statement

The authors declare that no conflict of interest exists as defined by the guidelines of the International Committee of Medical Journal Editors.

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ORIGINAL ARTICLE

Stable Incidence of Systemic Vasculitides in Schleswig-Holstein, Germany

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Schleswig-Holstein Vasculitis Registry, 1998-2005 (n=982)

- Wegener's granulomatosis (n=191)
- Microscopic polyangiitis (n=54)
- Churg-Strauss syndrome (n=28)
- Giant cell arteritis (n=272)
- Cutaneous leukocytoclastic angiitis (n=133)
- Henoch-Schönlein purpura (n=117)
- Unclassified vasculitis (n=135)
- Classic panarteritis nodosa (n=19)
- Takayasu's arteritis (n=9)
- Kawasaki syndrome (n=12)