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Incidence of ANCA-associated vasculitis in a UK mixed ethnicity population.

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Running title: Incidence of ANCA-associated vasculitis

Abstract

Objectives: We aimed to estimate the incidence of ANCA-associated vasculitis in the UK and how this varied by ethnic group.

Methods: We identified incident cases of ANCA-associated vasculitis between March 2007 and June 2013 in the Nottingham-Derby urban area from medical records using multiple sources. We derived the denominator population from the 2011 census, and we calculated incidence rate ratios using Poisson regression.

Results: Overall we identified 107 cases of ANCA-associated vasculitis, giving an incidence of 23.1/million person-years (95% CI 18.9-27.9). The incidence among the White population was 25.8/million person-years (95% CI 21.0–31.3), and among the Black and minority ethnic (BME) population was 8.4/million person-years (95% CI 3.1-18.3). After adjustment for age and sex, the difference between ethnic groups was not statistically significant (incidence rate ratio 0.7, 95% CI 0.3-1.5, p=0.3).

Conclusions: Overall the incidence of ANCA-associated vasculitis was similar to other epidemiological studies. Crude incidence rates were lower in the BME than White population but this was partly explained by the older age profile among the White compared to BME population.

Keywords:

Incidence

Epidemiology

Vasculitis

Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis

Granulomatosis with Polyangiitis

Microscopic Polyangiitis

Eosinophilic granulomatosis with polyangiitis

Ethnic Groups

Main text

Background:

The evidence base for the epidemiology of ANCA-associated vasculitis is limited. However, the incidence has been studied in a number of populations globally but these are predominantly White (1). Several studies have suggested that the occurrence of ANCA-associated vasculitis may be less in non-White ethnic groups, but there has not been a detailed study of the incidence in a multiethnic European population. There are no data on the incidence of ANCA-associated vasculitis in different ethnic groups relevant to the Black and minority ethnic (BME) populations in the UK. Such data are important for health services planning to ensure equal access to services for all ethnic groups, and to give insights into disease aetiology. The population of the Nottingham-Derby urban area is multiethnic, therefore, as part of an audit of service provision and outcome of patients with ANCA-associated vasculitis, we recorded the pre-existing data on the ethnicity of patients attending our vasculitis services.

The aim of this analysis was to calculate the incidence of ANCA-associated vasculitis in Nottingham and Derby, and how this varied by ethnic group.

Method:

We completed a retrospective case note review of all adults (aged > 16 years) with newly diagnosed ANCA-associated vasculitis who were resident within the Nottingham-Derby urban area during a 6.25 year

period (March 2007 – June 2013). We chose this start date because a new laboratory database was introduced in Nottingham in March 2007, and it was not possible to access laboratory results prior to this date. The stop date for patient inclusion was June 2013.

Study area and denominator population

The Nottingham and Derby Urban Areas are contiguous (Fig 1). Together they cover an area of 237 km^2. This combined area is geographically distinct from other cities, and people with ANCA-associated vasculitis are referred to either Nottingham University Hospitals NHS Trust or the Royal Derby Hospital hence this was a good location to capture an incident cohort. We verified the boundary by speaking to General Practitioners on the periphery, about their referral practices, and to rheumatologists in Mansfield (to the North) and Leicester (to the South) to confirm they did not see people with ANCA-associated vasculitis from within our catchment area. This study is limited to people who received healthcare at a national health service (NHS) hospital, however as NHS care is universal and free at the point of use, we expect this to include the overwhelming majority of people.

We used the results from the Office for National Statistics (ONS) 2011 census to provide the denominator population (2). This was available broken down by age, sex and ethnicity for each middle layer super output area (MSOA, an ONS sub-division of England with an average population

of around 7000). The index of multiple deprivation 2015 (IMD15) was used as a measure of social deprivation, but these data were not available at individual patient level, only by lower layer super output area (LSOA, an ONS sub-division of England, areas with about 1500 people) so a separate analysis was performed as these data could not be included as a variable in the main analysis.

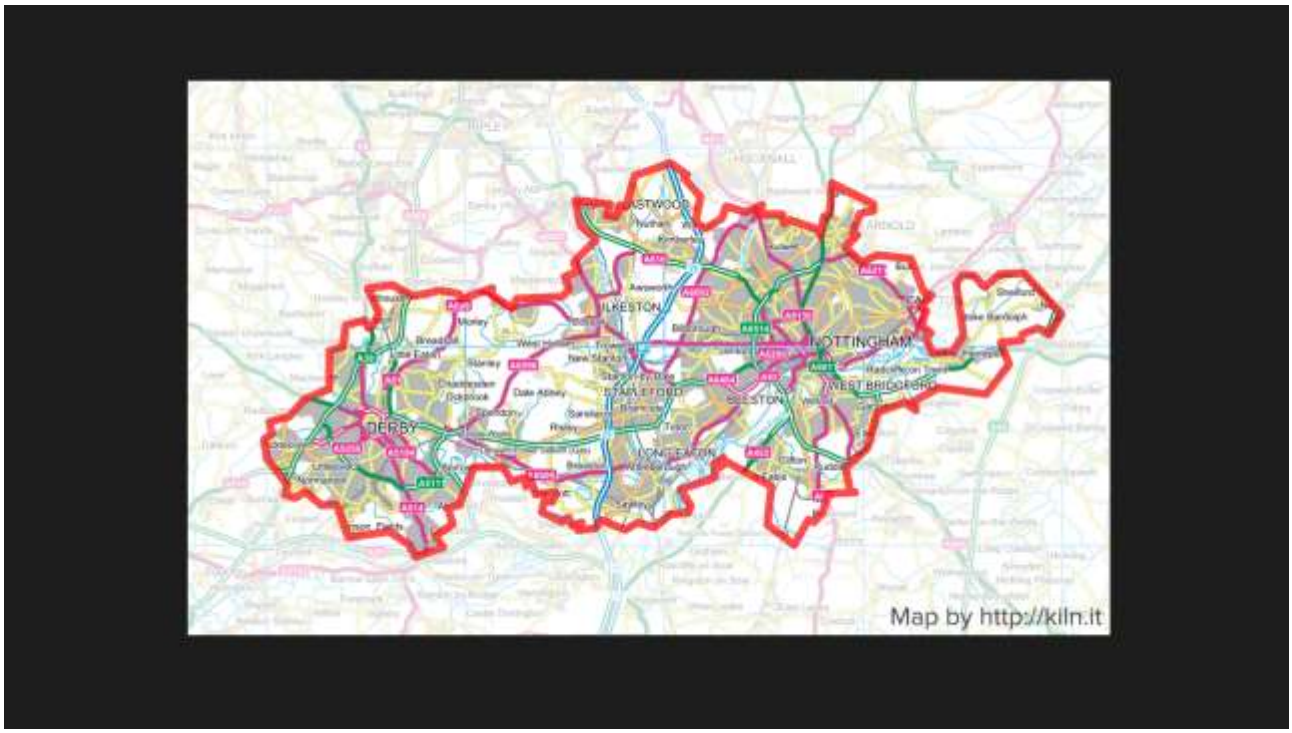

Figure 1 Study area: The continuous Nottingham and Derby urban areas. The area enclosed by the red line is the study population. (Map reproduced with permission from Kiln)

Sources of case ascertainment

Patients were identified from the hospital trust in Nottingham from 5 sources, 1) rheumatology departmental register, 2) renal departmental database search, 3) positive MPO/PR3 ANCA results, 4) histopathology laboratory database of biopsies coded for vasculitis (SnoMed codes), and 5) inpatient discharges coded as vasculitis using ICD 10 codes M301

polyarteritis with lung involvement (eosinophilic granulomatosis with polyangiitis [EGPA]-Churg Strauss), M313 granulomatosis with polyangiitis (GPA-Wegener's granulomatosis) and M317 microscopic polyangiitis (MPA). The study was carried out first in Nottingham, and as review of inpatient discharges and biopsies had only identified one extra patient we did not perform these additional reviews in Derby. Therefore, patients at the hospital in Derby were identified from 3 sources: 1) rheumatology department database search, 2) renal departmental database search and 3) positive MPO/PR3 ANCA results.

Case definition

People met the study definition of ANCA-associated vasculitis if they fulfilled 4 criteria: 1) new diagnosis of ANCA-associated vasculitis between 1 March 2007 and 30 June 2013, 2) residence within the study area at the time of diagnosis 3) positive ANCA by indirect immunofluorescence or ELISA, and 4) ANCA-associated vasculitis that was classifiable according to the EMA classification algorithm(3) which has been previously validated for use in epidemiological studies. This algorithm combines the Chapel Hill Consensus definitions for ANCA-associated vasculitis, standard disease classification criteria for GPA, MPA and EGPA, as well as including surrogate markers for upper airways granuloma and for renal vasculitis and ANCA. This was performed by FP. If there was uncertainty about diagnosis or classification, the case details were reviewed by PCL, RAW and Dr Al Ferraro (in Nottingham) or Dr J Leung (in Derby).

Data collection

In keeping with our audit protocol, data were extracted from the existing medical records on demographics (age, sex, ethnicity), postcode at time of diagnosis, clinical features sufficient to apply the EMA classification algorithm, ANCA status, and mortality. Postcode at time of diagnosis was used to determine residence within the study area, and to determine Middle layer super output area using the Office for National Statistics 'Neighbourhood statistics' website(4). Postcode was also used to look up the IMD15 quintile for the patient's Lower layer super output area (LSOA), where 1 denotes the most deprived 20% of LSOAs, and 5 the least deprived 20% of LSOAs. Ethnicity was ascertained from self-reported ethnicity recorded on the hospital patient information system using the 16+1 codes used by the health and social care information centre(5).

Data capture/recapture analysis was performed to estimate completeness of case ascertainment. This technique fits log-linear models to tables of the frequencies of which sources patients appeared in, and based on the overlap between the sources estimates the number of cases missed by all the sources and a confidence interval (6,7). This was done separately in Nottingham and Derby, as the sources were different.

To estimate the number of incident cases of ANCA-associated vasculitis in the UK in 2014, and projected numbers in 2024 and 2034 we used direct

standardization of the incidence rates calculated by age group to the ONS estimates for the UK population by age-group.

Statistical analyses

We categorised age into groups (16-39, 40-54, 55-69, 70-84 and 85+ years), and ethnicity into White, Black, Indo-Asian, Other Asian and Other/mixed ethnic groups. Ethnicity was also coded as a binary variable (White vs Black and minority ethnic [BME]) to increase statistical power. We calculated crude incidence rates (and Poisson 95% confidence intervals [95% CI]) per million person-years, and stratified these by age, ethnic group, and sex. Poisson regression modelling was applied firstly to one variable at a time, then all were included in a multi-variable model to control for age and sex. Due to the small size of this dataset, the Poisson regression model was re-run just for White versus all BME groups controlling for age and sex. We could not include socio-economic status in the main analysis as these data were not available at the individual patient level, instead they are only available as IMD-15 deprivation scores applied to each LSOA (areas of about 1500 people). The number of patients in each deprivation quintile was compared with the number expected in each quintile based on the deprivation scores (quintiles) assigned to all 567 LSOAs covered by the study area and the differences between these observed and expected values were compared using a chi-square test.

All analyses were performed using Stata 13 statistical software (Statacorp, Texas, USA)(8), except for the data capture/recapture analysis for a closed population which was performed using R-3.2.2 (R Core Team 2015), package RCapture(9).

Ethical aspects

This study was approved by the Nottingham University Hospitals NHS Trust and Royal Derby Hospitals NHS Trust clinical audit departments, as a service evaluation component of an audit against the BSR Guidelines for the Management of adults with ANCA-associated vasculitis, April 2013. (Nottingham University Hospitals NHS Trust audit department Project number 13-037C; Royal Derby Hospitals NHS Trust audit department project number RB-Rhe-2014/15-48).

Results:

The 2011 census adult (≥16 years) population within our defined area was 741071 adults (51% female). The ethnic breakdown for adults in this area was: 84.6% White and 15.4% BME (6.1% Indo-Asian, 2.4% other Asian, 3.4% Black, 3.5% mixed/ multiple/other ethnic groups).

In Nottingham we identified 1354 possible ANCA-associated vasculitis patients on the renal department register (50), rheumatology department register (54), positive MPO/PR3 ANCA results (812), pathology laboratory database of biopsies coded for vasculitis (207), renal biopsies with ANCA-associated vasculitis (87), and inpatient discharges coded as vasculitis

(144). Patients failed to fulfill the entry criteria for the following reasons: residence outside the study area at time of diagnosis, or diagnosis before or after the study timeframe, or no diagnosis of ANCA-associated vasculitis according to the EMA classification criteria, or a combination of these. Biopsy review and review of inpatient records only yielded one patient not identified by other means. Review of these 1354 records identified 64 patients who met the study inclusion criteria (see table 1). In Derby, we identified 572 possible patients on the renal department register (106), rheumatology department register (79), and positive MPO/PR3 ANCA results (387). Review of these 572 records identified 43 patients who met the study inclusion criteria (see table 1). The reason why some patients are included but do not appear on the list of positive MPO/PR3 results is that they were ANCA positive by indirect immunofluorescence only, and MPO/PR3 negative in our laboratory.

Overall we identified 107 incident cases of ANCA-associated vasculitis during the 6.25 year period in the denominator population. The demographic and clinical characteristics of the incident cases are shown in table 2. The majority of cases were men (60%) and median age at diagnosis was 70.2 (interquartile range [IQR] 58.4-78.6) years. 94.4% of cases were white, 1.9% were Black, 2.8% were Indo Asian, and 0.9% were other Asian. There were no cases among other or mixed ethnic groups. The overall annual incidence of ANCA-associated vasculitis was 23.1/million person-years (95% CI 18.9-27.9).

The incidence rates of GPA, MPA and EGPA were 8.2/million (95% CI 5.8–11.3), 13.4/million (95% CI 10.3–17.2) and 1.5/million (95% CI 0.6–3.1) respectively. The incidence of MPO-ANCA vasculitis was 11.9/million (95% CI 8.9-15.5) and PR3-ANCA vasculitis was 10.8/million (95% CI 8.0-14.2). There were a higher number of MPO+ patients (51.4%) compared to PR3+ patients (46.7%), and 3 patients (2.8%) were p-ANCA+ but MPO/PR3 negative.

Stratified crude incidence rates and incidence rate ratios (IRR) are presented in table 3. There was a marginal increase in the incidence in males compared to females (IRR 1.5, 95% CI 1.0-2.3]), and a dose-related increased incidence with increasing age, with the IRR for 40-55 age group compared to the 16-39 age group of 4.1 (95% CI 1.6-10.7) and increasing to 31.1 (95% CI 11.6-84.7) for the 85+ age category compared to 16-39 age group. Prior to adjustment, there was a significantly lower incidence of ANCA-associated vasculitis among the BME population compared to the White population (IRR 0.3, 95% CI 0.1-0.7]).

In the adjusted analysis (table 3b) the effect of ethnicity was reduced with IRR among BME people compared to White people of 0.7 (95% CI 0.3-1.5). Males had a higher risk of ANCA-associated vasculitis than females with an adjusted IRR of 1.8 (95% CI 1.2-2.6, p=0.004). Age had a large influence on incidence, with an IRR of 4.0 (1.5-10.4) for the 40-55

age group and 32.3 (95% CI 11.9-88.3) for the 85+ age group (reference group 16-39 years, p for trend<0.001).

A chi-squared test to compare the distribution of IMD15 among cases compared to the number expected based on the distribution of deprivation scores in the whole study area was not statistically significant (p=0.2).

The results of direct standardization of the incidence rates by age group from this study to the ONS estimates for the UK population by age-group, give the estimated number of cases of ANCA-associated vasculitis in the UK and are shown in table 5. The estimated number of incident cases in 2014 was 1348, predicting a rise to 1600 in 2024 and 1835 in 2034.

Capture/recapture analysis confirmed that overall there was a high degree of completeness of case capture within in our population (see table 4). In Nottingham the best-fitting model suggests we missed 0.3 cases (95% CI 0-1.5), and Derby 0.6 (95% CI 0-2.6) cases, giving an overall total of 0.9 missed cases. This best fitting model assumed independence between the three lists in their ability to identify cases.

Table 1: Numbers of patients on each list, and numbers included in the analysis

List	Patients on list with possible AAV (n)	Had a diagnosis of AAV & met the study geographical and time inclusion criteria (n)
Nottingham Rheumatology register	54	28
Nottingham Renal register	50	21
Nottingham Positive MPO/PR3	812	62
Nottingham Discharges	144	21
Nottingham Renal biopsies register	87	31
Nottingham Biopsies	207	4
Total in Nottingham	**1354**	**64**
Derby Rheumatology register	79	7
Derby Positive MPO/PR3	387	42
Derby Renal register	106	21
Total in Derby	**572**	**43**
Total Included		**107**

There were 2 patients with positive ANCA (by indirect immunofluorescence) but negative MPO/PR3 ANCA who were included in the study, which is why they did not appear on the positive MPO/PR3 lists. There was also 1 patient we missed due to human error.

Table 2: Demographics and clinical characteristics of the 107 incident cases

Demographics	
Female, n (%)	43 (40.2)
Male, n (%)	64 (59.8)
Median Age (yrs) (IQR)	70.2 (58.4-78.6)
Ethnicity, n (%)	
White	101 (94.4)
Black	2 (1.9)
Indo Asian	3 (2.8)
Other Asian	1 (0.9)
Other	0 (0)
Clinical Characteristics	
Diagnosis, n (%)	
EGPA	7 (6.5)
GPA	38 (35.5)
MPA	62 (57.9)
ANCA, n (%)	
PR3	50 (46.7)
MPO	55 (51.4)
IIF only	2 (1.9)
Active urinary sediment, n (%)	70 (65.4)
GN on renal biopsy, n (%)	63 (58.9)
ESRD, n (%)	29 (27.1)
Pulm Haemorrhage, n (%)	17 (15.9)
Dual GBM disease, n (%)	3 (2.8)
Death within 3 months, n (%)	14 (13.1)
Death within 1 year, n (%)	21 (19.6)

Active urinary sediment defined as ≥2+ blood & ≥2+ protein on urinalysis (3). GN: glomerulonephritis; ESRD: End Stage Renal Disease; Pulm Haemorrhage: pulmonary haemorrhage; GBM: Diagnosis of dual AAV and Anti-glomerular basement membrane (anti-GBM) disease.

Table 3: Incidence rates (5 Ethnicity categories)

	Cases	Denominator population	Crude incidence rates (95% CI) per million person-years
Ethnicity			
White	101	627103	25.8 (20.7-30.8)
BME	6	113968	8.4 (3.1-18.3)
Black	2	25231	12.7 (1.5-45.8)
Indo-Asian	3	45337	10.6 (2.2-30.9)
Other Asian	1	17537	9.1 (0.2-50.8)
Other	0	25863	0 (0-22.8)
Sex			
Female	43	376931	18.3 (13.2-24.6)
Male	64	364140	28.1(21.7-35.9)
Age group			
16-39 years	6	325168	3.0(1.1-6.4)
40-54 years	14	184233	12.2(6.6-20.4)
55-69 years	33	133053	39.7(27.3-55.7)
70-84 years	43	79578	86.4(62.5-116.5)
85+ years	11	19039	92.4(46.1-165.4)

Table 4: Incidence rates and Poisson regression modelling (2 Ethnicity categories)

	Cases	Denominator population	Unadjusted Rate ratios (95% CI) using single variable Poisson regression	Adjusted Rate ratios (95% CI) using multi-variable Poisson regression
Ethnicity				
White	101	627103	1.0	1.0
BME	6	113968	0.3(0.1-0.7)	0.7 (0.3-1.5) **P=0.3**
Sex				
Female	43	376931	1.0	1.0
Male	64	364140	1.5(1.0-2.3)	1.8 (1.2-2.6) **P=0.004**
Age group				
16-39 years	6	325168	1.0	1.0
40-54 years	14	184233	4.1(1.6-10.7)	4.0(1.5-10.4)
55-69 years	33	133053	13.4(5.6-32.1)	12.8(5.3-30.6
70-84 years	43	79578	29.3(12.5-68.6)	28.6(12.1-67.5)
85+ years	11	19039	31.3(11.6-84.7)	32.3(11.9-88.3) **P trend <0.001**
				Overall p<0.0001

Table 5: Data Capture-Recapture analysis using 3 sources

Nottingham									
Rheum/Renal list	+ve ANCA list	Biopsy/Discharge list	Frequency	model	Mean estimate of cases	SE	p	AIC	BIC
1	0	0	1	**c1+c2+c3**	**64.3**	**0.6**	**0.47**	**32.55**	**41.18**
1	1	0	17	c1+c2+c3+c1*c2	64.7	1.1	0.4	33.81	44.6
1	1	1	25	c1+c2+c3+c1*c3	64.3	0.6	0.29	34.5	45.29
1	0	1	0	c1+c2+c3+c2*c3	64.5	0.9	0.33	34.25	45.04
0	1	1	12	c1+c2+c3+c1*c2+c1*c3	64.7	1.1	0.18	35.79	48.75
0	1	0	8	c1+c2+c3+c1*c2+c2*c3	4851652095	2.04 e14	0.97	34.02	46.97
0	0	1	1	c1+c2+c3+c1*c3+c2*c3	64.5	0.9	0.14	36.22	49.17
				c1+c2+c3+c1*c2+c1*c3+c2*c3	4756521589	2 e14	1	36.02	51.13
Derby									
Rheum list	Renal list	+ve ANCA	Frequency	Model	Mean estimate of cases	SE	p	AIC	BIC
1	1	1	1	c1+c2+c3	43.8	1.2	0.19	29.88	36.92
1	1	0	0	**c1+c2+c3+c1*c2**	**43.6**	**1**	**0.74**	**27.68**	**36.49**
1	0	1	6	c1+c2+c3+c1*c3	44.1	1.6	0.13	31.19	40.00
1	0	0	0	c1+c2+c3+c2*c3	43	0	0.1	31.55	40.36
0	1	1	19	c1+c2+c3+c1*c2+c1*c3	43.8	1.3	1	29.07	39.64
0	1	0	1	c1+c2+c3+c1*c2+c2*c3	43	0	0.75	29.17	39.74
0	0	1	16	c1+c2+c3+c1*c3+c2*c3	44	13331.2	0.04	33.19	43.76
				c1+c2+c3+c1*c2+c1*c3+c2*c3	43.1	13825.3	1	31.07	43.40

Best-fitting model in bold typeface for Nottingham and Derby, indicated by having the lowest Akaike information criterion (AIC) and lowest Bayesian information criterion (BIC)(10).

Discussion

This study adds to the information on incidence of ANCA-associated vasculitis in the literature and is the first report to describe the incidence of ANCA-associated vasculitis in a multi-ethnic population from the UK. The main findings are that increasing age was strongly associated with increased disease incidence, and male sex marginally so. The crude incidence of ANCA-associated vasculitis was significantly lower in the BME population compared to the White population, however once adjusted for age and sex it was not statistically significant at a 5% level. This shows that the association between lower incidence of ANCA-associated vasculitis and BME ethnicities in the unadjusted analysis is partly explained by the age structure of the White population in Nottingham and Derby which is on average older than the BME population. The lack of statistical significance of the adjusted result may reflect in part the small size of this study, and a larger study with greater statistical power is needed to better estimate if there is a true difference in incidence between the ethnic groups or whether this occurred by chance. It was not possible to include social deprivation in the multivariable analysis, as data on the denominator population were available by ethnicity, age and sex, but not social deprivation. However, a chi-squared test of the distribution of IMD15 among cases compared to the whole study area showed no significant difference. Under the assumption of the incidence remaining stable over time, we estimate a 34% increase in the number of incident cases of ANCA-associated

vasculitis seen annually in the UK over the next 20 years, based on the predicted aging of the population.

The main strength of the study is robust case ascertainment of an incident cohort. The case ascertainment strategy was designed to minimise bias towards specific manifestations of ANCA-associated vasculitis – in particular the strategy of examining the case notes of all people with a positive MPO/PR3 ANCA or a biopsy (from any organ) which was coded as showing 'vasculitis'. This was in addition to the lists held by the Renal and Rheumatology departments which might omit predominantly ENT/neurological/respiratory cases, however, local practice in both hospitals in the study is for ENT/neurological and respiratory cases of ANCA-associated vasculitis to be referred to Rheumatology for administration of immunosuppression and hence would have been identified. The data capture/recapture analysis showed good completeness of case finding, however this does not estimate patients missed completely by the hospitals in the study, for example by dying before diagnosis, or by seeking their healthcare elsewhere. To address these issues we made efforts to include patients who died very soon after diagnosis, and our survival of 86.9% at 3 months (table 2) which is lower than previous published studies(10,11) suggests that we were successful. Also to address these issues, the boundaries of the study area were drawn conservatively close to the catchment hospitals to maximize the chance of patients being treated at those hospitals. Discussions with colleagues in neighbouring institutions

indicated that it was unlikely that patients from our catchment population would have been referred elsewhere. A review of the literature shows that methods for determining hospital catchment areas are historically contentious, and what is more, catchment areas may not be the same for different services based in the same hospital or Trust (12–15). However, the catchment area as defined by acute admissions in 2009 for Nottingham and Derby hospitals was 1,330, 624(16) and our population was only the 741,070 adults closest to the city centres, which provides some reassurance that the area we chose is conservative enough.

Inherent to all studies of rare diseases, the main weakness of this study is lack of power. Although the BME denominator population was 113967 and was followed for 6.25 years, there were only 6 incident cases of ANCA-associated vasculitis. Thus we were unable to determine the incidence rates for different minority ethnicities. The study was conducted across two hospital catchment areas to increase the size of the study population. The time-consuming nature of the methods of case ascertainment made it unfeasible to extend the study area further.

Overall our incidence figure is in line with that observed in a similar population based studies from 2005-2009 in Norfolk (UK) and in Japan(17) where the incidence of ANCA-associated vasculitis was 21.8 (95% CI 12.6-30.9) and 22.6 (95% CI 19.1-26.2) per million person-years, respectively. There have been

studies from Europe, Japan, USA, New Zealand and Canada using different methods of case ascertainment, and of case definition, reporting incidence rates of ANCA-associated vasculitis that range from 13 to 20/million person-years(1). The reason for our incidence rate being at the top end of this range is partly that incidence estimates increased during the 1990s when ANCA testing became routinely available, and also that we used a more stringent method of case ascertainment than most studies.

Unexpectedly, our study found the incidence of Microscopic polyangiitis (13.4/million, 95% CI 10.3–17.2) to be higher than of Granulomatosis with polyangiitis (8.2/million, 95% CI 5.8–11.3), and the incidence of MPO+ ANCA-associated vasculitis (11.9/million, 95% CI 8.9-15.5) to be higher than that of PR3+ ANCA-associated vasculitis (10.8/million, 95% CI 8.0-14.2). In comparison to the previous study by Watts et al(18) in Norfolk which found the incidence of GPA to be 11.3, 9.1-13.4) and MPA to be 5.9 (4.4-7.5), the two studies used the same case definitions and the same strategy of retrospective case note review. However there were differences in case-finding strategies – the Norfolk study did not ascertain cases from all positive MPO/PR3 ANCA tests. Our results are most similar to Mohammad et al's study from Sweden which used a similar strategy of case ascertainment to our study and estimated the overall incidence of AAV to be 20.9 (17.3 – 24.4), and the incidence of MPA (10.1, 95% CI 7.7-12.6) to be higher than the incidence of

GPA (9.8, 95% CI 7.4-12.2)(19). As yet undefined environmental factors may also contribute to these differences.

It is difficult to compare the findings of our incidence study to studies of prevalence, as prevalence is influenced by survival as well as incidence. However, it is interesting to note the previous observation that the prevalence of ANCA-associated vasculitis in a multi-ethnic population of the greater metropolitan Paris area was two-fold higher in the subjects of European ancestry compared to the non-Europeans(20) would be compatible with our study. Ignoring any possible differences in survival between ethnic groups, which would affect prevalence, our unadjusted incidence rate for Whites compared to BME was about triple.

In this paper we are not able to comment on whether differences in risk of disease in people with different self-reported ethnicities relate to their racial (genetic) origins or social and cultural practices. Recent data suggests that the White population of central England is homogeneous and is of Northern European Caucasian race (21) and our overall incidence is comparable with incidence figures from other studies in Northern European populations(1).

This study estimates projected UK numbers of incident cases for future planning of services for patients with ANCA-associated vasculitis. The strong association between increasing age and increased incidence of ANCA-

associated vasculitis found in this study suggests that the number of cases requiring treatment is likely to increase as the population ages, and we predict a 34% increase in the number incident cases annually in the UK over the next 20 years if our study area is representative of the UK. It is important to note that this projection is based solely on anticipated changes to the age demographic and will not account for changes in environmental influences which may be important (e.g. dust exposure) and which may also change throughout this time period. Increasing numbers of patients from BME communities may be seen if the BME age structure becomes more similar to the White population.

To conclude, the overall incidence of ANCA-associated vasculitis in Nottingham/Derby UK was similar to other epidemiological studies in the UK and worldwide. Further work to confirm these findings and explore the relationship with ethnicity further should be conducted in larger datasets, for example using the UK databases of routinely collected healthcare data, which have routinely collected ethnicity data since 1991(22,23).

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Legends for illustrations:

Figure 1

Study area: The contiguous Nottingham and Derby urban areas. The area enclosed by the red line is the study population.

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Key messages:

1. The incidence of ANCA-associated vasculitis in the Midlands of the UK was estimated at 23.1/million person-years.
2. The age-and-sex-adjusted incidence in the BME and White populations was similar.
3. The annual number of new cases of ANCA-associated vasculitis in the UK is estimated to increase by 34% over the next 20 years due to the aging population.