
Supplementary information

Global epidemiology of vasculitis

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Supplementary Tables.

Supplementary Table 1. Incidence of giant cell arteritis.

Region	Study period	Incidence (cases per 100,000 population aged > 50 year unless specified)	Case definition	Study type	Reference
<i>Europe</i>					
Denmark	1996-2018	22.2	ICD-10 codes DM31.5 and DM 31.6)	Administrative database analysis	¹
Savo, Finland	2010	7.4 (female 10.6, male 4.9)	ACR 1990 criteria	Population Survey	²
France	2007-2015	10.0	ICD-10 codes (DM31.5 and DM 31.6)	Administrative insurance database analysis, retrospective	³
Germany	1998-2002	3.2	CHCC 1994	Prospective population survey	⁴
Reykjavik, Iceland	1967-2008	43.6	Biopsy positive	Population based cohort study	⁵
Jerusalem, Israel	1990-2009	8.1 (Jewish population)	ACR 1990 criteria /CHCC 2012	Hospital based retrospective, ACR or CHCC	⁶
Reggio Emilia, Italy	1986-2012	5.8 (females 7.8, males 3.3)	Biopsy positive	Analysis pathology database	⁷
Southern Norway	2000-2013	16.8 (females 24.5, males 10.2)	ACR 1990 criteria/Biopsy/imaging	Hospital data base analysis using ICD DM31.5 and DM 31.6	⁸
Slovenia	2012-2017	8.7 (females 120.6, males 6.4)	ACR 1990 criteria & biopsy positive	Prospective population based	⁹
Spain	1981-2005	10.1 (females 10.2, males 9.9)	Biopsy positive	Retrospective case note review	¹⁰

Sweden	1997-2019	13.3 (females 17.8, males 8.2)	Biopsy positive	Analysis pathology database	¹¹
Turkey	2003-2009	1.1 (females 1.14, males 1.1)	ACR 1990 criteria	Hospital based	¹²
United Kingdom	2000-2011	10.0 (females 15, males 6.0)	Read codes (G75500, G755.00, G755z00, N200.00, G755100), age >50 years, steroids within 6 months of registry entry	Analysis clinical population database (UK CPRD)	¹³
<i>North America</i>					
Olmsted county, USA	2000-2009	19.8 (females 27.0, males 10.1)	ACR 1990 criteria	Population based cohort	¹⁴
Saskatoon, Canada	1998-2003	9.4 (including Europeans and Indigenous) 7.7 Europeans 16 Indigenous	Biopsy positive	Analysis neuro-ophthalmology database	¹⁵
<i>Australasia</i>					
South Australia	1992-2011	3.2 (females 4.3, males 1.9)	Biopsy positive	Analysis pathology database	¹⁶
Otago, New Zealand	1996-2005	12.7	Biopsy positive	Analysis pathology database	¹⁷

ACR – American College Rheumatology, CHCC – Chapel Hill Consensus Conference, ICD – International Classification of Diseases
ACR 1990 criteria for GCA¹⁸; CHCC 1994¹⁹

Supplementary Table 2. Incidence of Kawasaki disease.

Region	Study period	Incidence (cases per 100,000 population aged > 5 year unless specified)	Case definition	Study type	Reference
<i>Europe</i>					
Czech Republic	1997-1999	1.6	AHA	Hospital surveillance	²⁰
Denmark	1981-2004	4.9	Clinical	Analysis hospital discharge records	²¹
Finland	1999-2009	11.4	ICD 9 code 446B or 446.98 or ICD 10 code M30.3	Analysis hospital discharge database	²²
Northern France	2005-2006	9.0	AHA	Cohort study	²³
Germany	2011-2012	7.2	AHA	Nationwide surveillance	²⁴
Georgia	1997-1998	9.8	CDC	National hospital discharge database	²⁵
Greece	2001-2010	3.2	AHA	Hospital discharge database analysis	²⁶
Ireland	1996-2000	15.2	Clinical	National hospital discharge analysis	²⁷
Israel	1996-2009	6.4	ICD9 446.1	Analysis national hospital discharge register	²⁸
Italy	2008-2013	17.6	ICD9 446.1	Regional hospital discharge database analysis	²⁹

Norway	1999-2009	5.4	ICD 9 code 446B or 446.98 or ICD 10 code M30.3	Analysis hospital discharge database	²²
Spain	2005-2015	11.7	ICD -9 446.1	Analysis of hospital morbidity survey	³⁰
Sweden	1999-2009	7.4	ICD 9 code 446B or 446.98 or ICD 10 code M30.3	Analysis hospital discharge database	²²
The Netherlands	2008-2012	5.8	Clinical	Prospective surveillance	³¹
United Kingdom	2014-2015	4.5	AHA	Population surveillance	³²
<i>Americas</i>					
Ontario, Canada	2000-2014	19.6	Discharge diagnosis	Analysis National hospital discharge database	³³
USA	2018	19.8	ICD-CM	Analysis administrative database	³⁴
Panama	2009-2013	2.1	AHA 2004	Retrospective review national	³⁵
<i>Australasia</i>					
Australia	2000-2009	9.3	AHA	Analysis discharge database	³⁶
New Zealand	2001-2002	8.0	AHA	National surveillance	³⁷
<i>Asia</i>					
China	2000-2004	55	Clinical	Hospital based survey	³⁸
Hong Kong	2000-2011	74	AHA		³⁹
India	1994-2008	5.35	AHA	Hospital database analysis	⁴⁰
Japan	2017-2018	359	Japanese guidelines	Nationwide survey	⁴¹

South Korea	2015-2017	197	Clinical	Population survey	⁴²
Taiwan	1996-2011	45.2	ICD-9-CM 4461	Nationwide retrospective. Analysis insurance database	⁴³
Thailand	1998-2002	3.4	AHA	National registry	⁴⁴
Malaysia	2006-2009	14.8	AHA	Population	⁴⁵

AHA = American Heart Association⁴⁶

Supplementary Table 3. Incidence of ANCA-associated vasculitis (AAV).

Region	Study Period	Incidence (cases per million population)			Case Definition	Study type	Reference
		GPA	MPA	EGPA			
<i>Europe</i>							
Nottingham, UK	2007–2013	8.2	13.4	1.5	EMA	Population register	⁴⁷
Tromso, Norway	1999–2013	15.6	10.4	2.7	EMA	Population register	⁴⁸
Lund, South Sweden	1997–2006	9.8	10.1	0.9	EMA	Population register	⁴⁹
Finland	1996–2000	9.3	NA	NA	GPA ACR 1990	Analysis national discharge data	⁵⁰
Germany	2013–2016	34.0	13.0	NA	GPA ICD-10 M31.3 MPA ICD-10 M31.7	Analysis insurance claims database	⁵¹
Vilnius, Lithuania	1990–1999	2.1	3	1.3	GPA ACR 1990 MPA ACR 1990 for PAN EGPA ACR 1990	Analysis hospital database	⁵²
Crete, Greece	1995–2003	6.6	10.2	NA	GPA ACR 1990 MPA CHCC 1994	Analysis hospital records	⁵³
Malaga, Spain	1994–2010	2.1	3.4	0.64	GPA ACR 1990	Retrospective population	⁵⁴

					MPA CHCC 1994 EGPA ACR 1990		
Reggio Emilia, Italy	2004– 2009	3.4	NA	NA	ACR	Analysis hospital discharge database	⁷
Faroes (Danes)	1992– 2011	6.4 (Danes) 1.0 (Inuit)	NA	NA	GPA ACR 1990	Analysis national hospital register	⁵⁵
Burgundy, France	1998– 2008	NA	NA	1.2	EGPA ACR 1990	Analysis hospital discharge database	⁵⁶
Edirne, Turkey	2004– 2014	4.8	2.4	0.8	GPA ACR 1990 MPA CHCC 1994 EGPA ACR 1990	Retrospective analysis hospital database	⁵⁷
Jerusalem (Jewish population— hospital inpatients only)	1990– 2009	4.1	2.3	1.2	GPA ACR 1990 MPA CHCC 1994 EGPA ACR 1990	Analysis of hospital records. Jewish population only	⁶
Poland	2008– 2013	NA	NA	1.5	ICD-10	Analysis national hospital discharge database	⁵⁸

Poland	2011-2015	7.7	NA	NA	ICD-10	Analysis national hospital discharge database	⁵⁸
<i>Americas</i>							
Olmsted County, USA	1996-2015	13.0	16.0	4.0	GPA ACR 1990- MPA CHCC 2012 EGPA ACR 1990	Population register	⁵⁹
Saskatchewan, Canada [†]	2007–2011	4.6	7.1	NA	GPA ACR 1990 MPA CHCC 1994	Analysis hospital database	⁶⁰
Lima, Peru	1990–2004	0.5	4	0.14	CHCC	Hospital based population	⁶¹
Argentina	2000-2015	9	14	NA	GPA ACR 1990 MPA CHCC 2012	Analysis insurance database	⁶²
<i>Australasia</i>							
Capital Territory, + SE NSW, Australia	2000–2004	8.4	5.0	2.2	GPA ACR 1990 MPA CHCC 1994 ACR 1990	Analysis hospital inpatient database	⁶³
New Zealand	1999–2003	10.4	NA	NA	ICD 10 M301, M313	National hospital database	⁶⁴
<i>Asia</i>							

Japan	2005– 2009	2.1	18.2	NA	GPA ACR 1990 MPA CHCC 1994	Population register	⁶⁵
Taiwan	1997– 2008	0.37	NA	NA	GPA ICD-9 446.4	Analysis national health insurance database	⁶⁶
<i>Middle East</i>							
Kuwait	1993– 1996	NA	24.0	NA	MPA CHCC 1994		⁶⁷

[†]Renal involvement only; reported annual incidence of granulomatosis with polyangiitis (Wegener's) and microscopic polyangiitis. *ACR*, American College of Rheumatology criteria (1990); *CHCC*, Chapel Hill Consensus Conference definition (1994); *EGPA*, eosinophilic granulomatosis with polyangiitis; *EMA*, European Medicines Agency Algorithm; *GN*, glomerulonephritis; *GPA*, granulomatosis with polyangiitis; *ICD*, International Classification of Diseases; *MPA*, microscopic polyangiitis; *NA*, not available; *UKGPRD*, United Kingdom General Practice Research Database. ACR 1990 criteria for GPA⁶⁸, ACR 1990 criteria for EGPA⁶⁹, CHCC 1994 definitions¹⁹, CHCC 2012 definitions⁷⁰

Supplementary Table 4. Prevalence of Behcet syndrome.

Region	Study period	Prevalence (cases per 100,000 population)	Case definition	Study type	Reference
<i>Europe</i>					
Paris, France	2003	7.1	ISG 1990	Population	⁷¹
Germany	2012	1.5	ISG 2014	Analysis national registry	⁷²
Mid-west, Ireland	2017	6.2	ISG 2014	Cohort study	⁷³
Italy	2010	15.9	ISG 1990	Cross sectional	⁷⁴
Rotterdam, Netherlands	2012	7.6	ISG 1990	Analysis hospital database	⁷⁵
Poland	2014	0.34	ICD-10	Analysis national administrative database	⁷⁶
Portugal	1997	2.4	NA	Cohort study	⁷⁷
Spain	1988-1997	0.66	ISG 1990	Cohort study	⁷⁸
Southern Sweden	2011	4.9	ISG 1990	Population	⁷⁹
Switzerland	Not stated	4.0	ISGBD 2014	Cohort study	⁸⁰
United Kingdom	1995-2017	14.6	Read codes	National clinical database primary care	⁸¹
<i>Middle East</i>					
Egypt	2017	3.6	ISGBD 2014	Cross sectional	⁸²
Iraq	1999-2000	17	ISG 1990	Cross sectional survey	⁸³
Kurdistan, Iran	2011-2012	100	ISG 1990	Cross sectional population survey	⁸⁴
Israel	Not stated	120	ISG 1990	Population	⁸⁵
Jordan	Not stated	660	ISG	Cross sectional hospital workers	⁸⁶
Kuwait	N/A	2.1	N/A	Cohort study	⁸⁷

Lebanon	2007	100	ISG 1990	Cross sectional population survey	⁸⁸
Saudi Arabia	1979-1992	20	ISG 1990	Cohort study	⁸⁹
Istanbul, Turkey	Not stated	421	ISG 1990	Cross sectional population	⁹⁰
<i>Americas</i>					
Brazil	2006-2007	0.3	ISG 1990	Cross sectional hospital	⁹¹
Colombia	2012-2016	1.1	ICD-10 code: M352	Analysis administrative database	⁹²
South western, USA	Not stated	10.6	ISG 1990	Cross sectional hospital database	⁹³
<i>Asia</i>					
China	Not stated	10	ISG 1990	Cross sectional population survey	⁹⁴
Japan	Not stated	13.5	Not stated	Not stated	⁹⁵
Mongolia	2015-2019	2.4	ISG 1990	Cross sectional	⁹⁶
Korea	2011-2017	15.1	Korean Disease Classification	Analysis insurance database	⁹⁷
Taiwan	2000	1.4	ICD-9-CM 136.1	Analysis national insurance database	⁹⁸

ISG = International study group 1990

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