nature reviews rheumatology

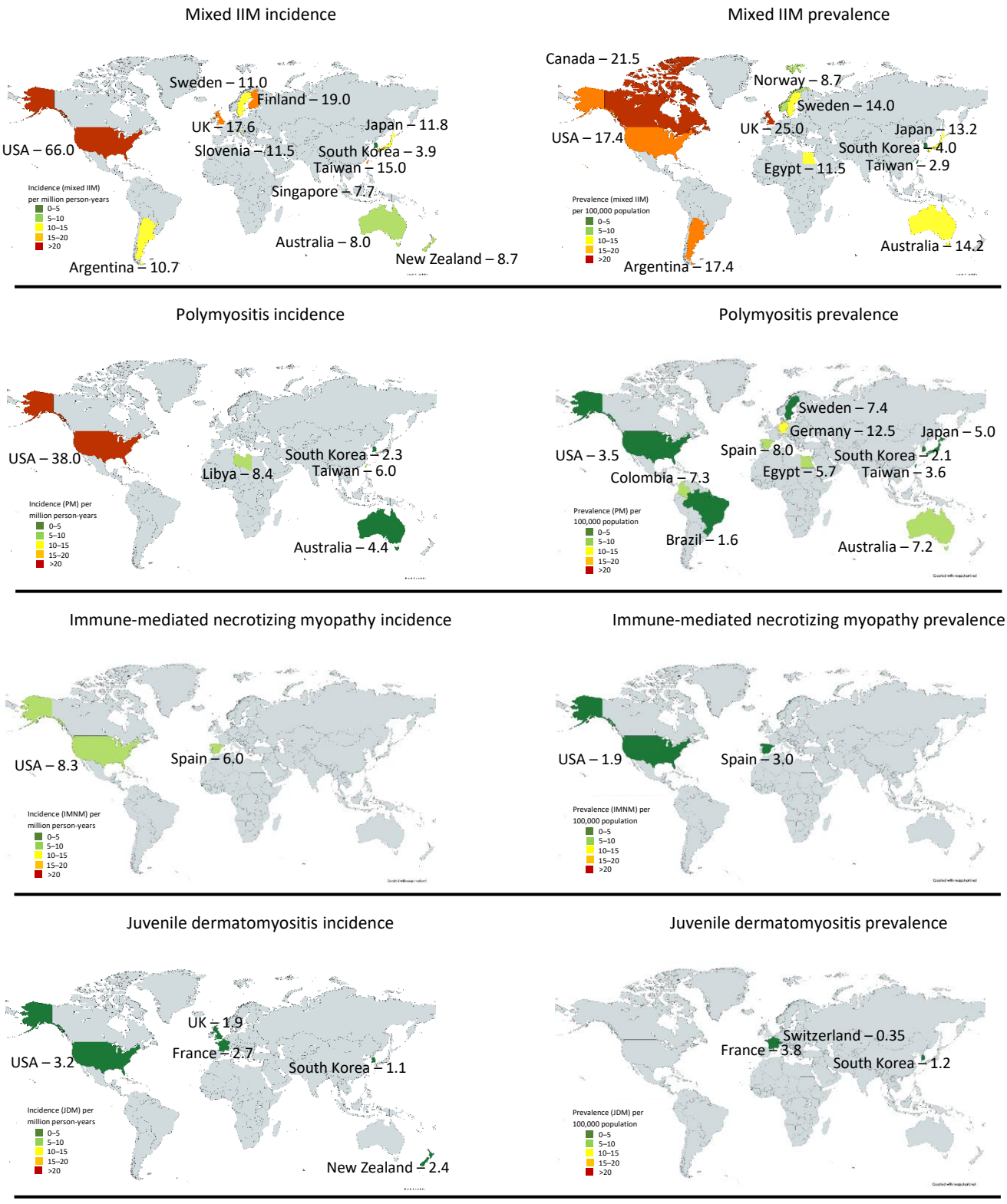
https://doi.org/10.1038/s41584-023-01033-0

Review article

Check for updates

Epidemiology of the idiopathic inflammatory myopathies

In the format provided by the authors and unedited



Supplementary Figure 1 | World maps showing global incidence per million person-years (left) and prevalence per 100,000 people (right) of: mixed cohorts of the idiopathic inflammatory myopathies (IIM), polymyositis (PM), immune-mediated necrotizing myopathy (IMNM) and juvenile dermatomyositis (JDM) from epidemiological studies which can be found in more detail in Supplementary Table 1. Countries with no published data are shaded grey. Similar maps for dermatomyositis and sporadic inclusion-body myositis are shown in Figure 3.

Supplementary Table 1 | Publications on the epidemiology of idiopathic inflammatory myopathies that report a disease incidence and/or prevalence

Region	Study description and time period	Population and subtypes of IIM	Demographics (ethnicity, age, sex (%))	Criteria for IIM	Incidence (per 100,000 person-years except where noted)	Prevalence (per 100,000 people except where noted)	Ref.
Mixed IIM subtypes							
North America							
USA	Review of nationally managed care organisation consisting of commercially insured members (2003–2008)	1,941 with IIM for the incidence cohort (396 – DM, 1,069 – PM, 559 – interstitial myositis)	Ethnicity not reported Median age 49 years 64.9% female (66.8% in the prevalence cohort)	ICD-9 coding: 710.3 (DM), 710.4 (PM), 728.81 (interstitial myositis)	6.6 – adjusted for age and gender (1.4 – DM, 3.8 – PM, 1.7 – interstitial myositis)	14 (2003) and 17.4 (2008) 23.7 female vs 10.6 male, 2008 Prevalence increases with age	[1]
USA	Retrospective review of commercial database covering outpatient medical and pharmacy claims data for individuals <65 years covered by private-sector health plans using ICD codes for people with IIM (2004–2008)	1,784 incident PM 1,072 incident DM 476 incident interstitial myositis	37.6% white 42.9% African American 4.9% Hispanic Most common age category was 55-64 years old 77.4% female	ICD-9 coding	4.27 (1.52 – DM, 2.46 – PM, 0.73 – interstitial myositis) adjusted for age and sex	18.5-20.4 (2004), 17.7-19.2 (2008) after sensitivity analysis	[2]
Alberta, Canada	Retrospective review of Alberta Health Care Insurance Plan (AHCIP), a universal publicly funded system, for cases of IIM. Cases defined by coding - ≥1 billing code by rheumatologist, ≥2 by any physician, or 1	Not reported	3.7% identified as First Nations Age not reported Sex not reported	ICD-9-CM coding for PM or DM	N/A	25 (42.6 – female, 6.5 - male) (First Nations) 33.8 (47.7 – female, 19.6 – male) (non-First Nations) Significantly higher prevalence in	[3]

	hospitalisation discharge summary (1994-2007)					rural vs urban dwellers e.g. 124.5 vs 86.7 for non-First Nations women >45-years old	
Quebec, Canada	Retrospective review of physician billing and hospitalisation databases covering the province using ICD codes to identify people with PM/DM (1989–2003)	Absolute numbers not reported	Ethnicity not reported Mean age 60 years (SD 18 years) 68.5% female	ICD codes	N/A	21.5 Prevalence lowest in young rural men (2.7) and highest in older urban women (70)	[4]
Olmsted County, USA	Retrospective review of hospital records from Mayo Clinic for patients with a diagnosis of 'myositis'. Subsequent review of clinical record to apply classification criteria (1981–2000)	9 with IBM (5 definite, 4 possible) 6 with PM	100% white Median age of PM: 59 years (range 23-86 years), IBM: 68 years (55-84 years) 76.5% female (66.7% for IBM subgroup)	Bohan and Peter Griggs	0.41 – PM, 0.79 – IBM (age and sex adjusted)	3.45 – PM, 7.06 – IBM (age and sex adjusted)	[5]
Shelby County, USA	Retrospective review of hospital records for discharge diagnoses of polymyositis, myositis or myopathy, and dermatomyositis (1947–1968)	124 with PM (considered to include DM at this time)	56.5% white 43.5% non-white (ethnicities within this group not reported)	ICD codes	0.5 (age adjusted) Increase in incidence with time (1.2 – 1947-1952, 8.4 – 1963-1968)	N/A	[6]
South America							
Colombia	Retrospective review of the Comprehensive Social Protection Information	3,507 with PM 7,277 with DM	Ethnicity not reported Median age in 35-39 range	ICD-10 coding: M33.0 (JDM), M33.1 (other DM), M33.2 (PM), M33.9	N/A	7.3 (PM) 15.1 (DM) Highest prevalence in	[7]

	System (SISPRO) which covers 95% of the population's health care records (2012–2018)		77% female	(DM/PM not otherwise specified)		60–69 years age group	
Buenos Aires, Argentina	Retrospective review of records of Hospital Italiano Medical Care Program (HIMCP), a private health insurance system, for a diagnosis of myositis or a related ICD-9 code with subsequent review applying classification criteria (1999-2009)	7 with PM 10 with DM 10 incident IIM cases (1999-2009): 7 PM, 3 DM	Ethnicity not reported Mean age at diagnosis 55 years (SD 15 years) 76.5% female	Bohan and Peter	1.07 (1.24 – female, 0.8 – male)	17.4 (22.1 – female, 10.2 – male)	[8]
Europe							
Germany	Retrospective review of outpatient and inpatient data from health insurance company using ICD codes to identify myositis. (2005-2019)	45800 with 'myositis'	Ethnicity not reported Age not reported Sex not reported	ICD-10 coding:	2.6 – PM (2009) 1.5 – PM (2019)	2019: 64.6 – total 11.3 – DM 12.5 – PM Higher prevalence in females (74.4 vs 51.7 in males, 2019)	[9]
UK	Retrospective review of Clinical Practice Research Datalink (CPRD) primary care database for neuromuscular conditions (2000-2019)	Various neuromuscular disease, 3,152 with IIM. 790 incident cases of IIM.	Ethnicity not reported Age and sex not reported for separate diagnoses	Not reported	1.3 (1.6 – female, -0.98 – male)	25 (31.3 – female, 18.6 – male) Highest prevalence in 75–79 years age group	[10]
Catalonia, Spain	Retrospective review of the Public Data Analysis for Health	575 with PM 1,458 with DM	Ethnicity not reported	ICD-9 coding	N/A	8 (PM) 19 (DM)	[11]

	Research and Innovation Program (PADRIS) using ICD-9 codes to identify a variety of autoimmune diseases (2012–2017)		Age not reported 62% female				
Ljubljana and Gorenjska, Slovenia	Retrospective review of medical records using ICD codes combined with muscle biopsy histopathology reports to identify incident cases of IIM (2010–2017)	18 incident DM 14 incident ASyS 11 incident overlap myositis 9 incident statin-associated IMNM 5 incident PM 4 incident CAM 2 incident anti-SRP IMNM 1 incident IBM 1 incident undifferentiated myositis	Ethnicity not reported Median age 64.8 years (range 22-91 years) 72.3% female	ICD codes and histopathological criteria	1.15 (1.6 – female, 0.66 – male) Incidence in females (but not males) increased with age	N/A	[12]
Salford, UK	Retrospective review of medical records from tertiary referral centre (Salford Royal NHS) neuromuscular clinic for adults diagnosed with IIM, using ICD-10 codes, clinic referrals and the UKMYONET database (2007–2016)	32 incident cases of IIM: 10 with PM, 8 with DM, 2 with IBM, 1 with CADM (by EULAR/ACR criteria)	Ethnicity not reported Mean age at onset 58.1 years (SD 14.2 years) 71.9% female	ICD-10 coding EULAR/ACR 2017 Expert opinion from two investigators	1.76 (2.52 – females, 1.0 – males) Increase in incidence with time: 1.36 (2007-2011) 2.14 (2012-2016)	N/A	[13]
Sweden	Retrospective review of the National Patient Register (NPR) for ICD codes of IIM, and of the	522 incident IIM (2007-2011): 267 with PM, 148 with DM, 53 with IBM, 36 with JDM	Ethnicity not reported Median age 63 years (IQR 49-72 years)	ICD-10 coding	1.1 (1.3 – female, 0.97 – male) Incidence increases	14 (17 – female, 11 – male) Peak prevalence in	[14]

	Swedish Quality of Care Register (SRQ) for possible IIM (2001–2012)	1,267 with IIM (2012): 698 with PM, 354 with DM, 90 with IBM, 102 with JDM	of prevalence cohort 61% female		with age. No difference with north-south gradient or urban-rural distinction.	70–79-year-old group (42)	
North Savo, Finland	Prospective data collection of new diagnoses of autoimmune connective tissue disease in five hospitals with rheumatology services (2010)	Various autoimmune CTD, 4 with IIM (2 with PM, 2 with DM)	Homogeneous white population Mean age 68.7 years (SD 10.4 years) 50% female	Bohan and Peter	1.9 (1.9 – female, 2.0 – male)	N/A	[15]
Southeast Norway	Retrospective review of patient administrative databases for ICD-10 codes with subsequent chart review of these patients to ensure meeting criteria (2003–2012)	100 with PM 130 with DM	Ethnicity not reported Mean age at diagnosis 53.5 years 65.2% female DM patients younger at diagnosis (51.6 vs 56.0 years, p = 0.03)	Targoff	N/A	8.7 Variation noted around the counties of SE Norway (highest in Telemark – 16.4)	[16]
Denmark	Retrospective review of the Danish Civil Registration System which is linked to the National Hospital Register which uses ICD8-10 codes (2001)	45,051 with 'myositis' 949 with 'dermato-polymyositis'	Ethnicity not reported Age not reported Sex not reported	ICD 8/10 (716/M33 – dermato-polymyositis, 717.90/M60 – myositis)	N/A	170 (dermato-polymyositis) 8,230 (myositis)	[17]
Gävleborg, Sweden	Retrospective review of all suggestive muscle biopsies in addition to ICD code searching of medical records	5 with PM 3 with DM 7 with overlap myositis	Ethnicity not reported Mean age 54 years (range 18–85 years)	Bohan and Peter Histopathological criteria for IBM	0.76	N/A	[18]

	with subsequent re-review of case to apply classification criteria (1984–1993)	3 with CAM 3 with IBM	52.4% female				
Five districts of Finland (Jyväskylä, Kotka, Kuopio, Lahti and Tampere)	Review of records in the database of the Sickness Insurance Act, which subsidises medication costs, for rare systemic rheumatic and connective tissue diseases (1990)	Various autoimmune rheumatic diseases, 5 with IIM (4 with PM or DM, 1 with sIBM)	Ethnicity not reported Age not reported Sex not reported	Bohan and Peter	0.37	N/A	[19]
Örebro, Sweden	Retrospective review of medical records, the National Health Insurance (NHI) register to identify people with various neuromuscular diseases by ICD codes (1988)	20 with PM 9 with DM	Ethnicity not reported Mean age overall 56 years (range 14–82 years), PM 61 years (17–82 years), DM 44 years (14–67 years) 56% female (67% for DM, 50% for PM)	ICD-8 and ICD-9 coding	N/A	11 (3.3 – DM, 7.4 – PM)	[20]
Asia							
South Korea	Retrospective review of the Korean National Health Insurance Service (NHIS) database, which includes >96.3% of the population, for IIM (2004–2015)	2,100 with IIM (130 JDM, 1,131 DM, 949 PM) in 2015	Ethnicity not reported Mean age 51.2 years (SD 16.9 years) 72.1% female	ICD-10 code and registration in the Individual Copayment Beneficiaries Program (ICBP) which utilizes Bohan and Peter criteria	0.44 (0.15 – JDM, 0.27 – DM, 0.28 – PM) in 2006 0.37 (0.07 – JDM, 0.2 – DM, 0.2 – PM) in 2015	2.3 (0.9 – JDM, 1.4 – DM, 1.4 – PM) in 2006 4.0 (1.2 – JDM, 2.7 – DM, 2.1 – PM) in 2015	[21]
Japan	Retrospective review of national registration system for intractable diseases to search	13,163 (2003) to 16,926 (2010) with PM/DM	Ethnicity not reported Age not reported	Tanimoto	0.68 (2003) 1.18 (2010)	10.3 (2003) 13.2 (2010)	[22]

	for patients with PM/DM (2003–2010)		Sex not reported				
Taiwan	Review of patients who have a Critical Illness Certificate for various autoimmune rheumatic diseases as part of the compulsory National Health Insurance System using ICD codes (2005–2009)	Various autoimmune rheumatic diseases, including: 36 with PM 24 with DM	Ethnicity not reported Age not reported Sex not reported	ICD-9 coding: 710.3 (DM), 710.4 (PM)	DM: 0.7 (0.8 – female, 0.6 – male) PM: 0.6 (0.8 – female, 0.6 – male)	DM: 2.4 (3.4 – female, 1.4 – male) PM: 3.6 (4.8 – female, 2.4 – male)	[23]
Taiwan	Retrospective review of the National Health Insurance Database (NIHRD) and National Death Registry (NHD) using ICD codes to identify various autoimmune diseases (2000–2008)	Various autoimmune rheumatic disease, 28 with PM/DM	Ethnicity not reported Age not reported 71.4% female	ICD-9 coding: 710.3 (DM), 710.4 (PM)	1.5 (1.9 – female, 1.0 – male)	2.9 (4.2 – female, 1.6 – male)	[24]
Taiwan	Review of patients who have a Critical Illness Certificate for PM/DM as part of the compulsory National Health Insurance System (2003–2007)	500 with PM 803 with DM	Ethnicity not reported Mean age at diagnosis 44 years (SD 18.3 years) – DM, 49.2 years (SD 15.9 years) – PM 69.7% female	ICD-9 coding	0.71 – DM 0.44 – PM 1.2 – female, 0.52 – male Incidence increased with age to peak in 50–59 years	N/A	[25]
Tottori Prefecture, Japan	Review of medical records combined with questionnaires to physicians for inflammatory neuromuscular conditions	61 with PM/DM with 31 incident cases	Ethnicity not reported Mean age at onset 52.6 years (SD 21.3 years)	Bohan and Peter	1.01 (0.48 – 1991, 1.94 – 1990) No geographical difference in incidence in	9.9 (13.7 – female, 5.8 – male)	[26]

	(1988–1992)		72.1% female		the prefecture		
Singapore	Retrospective review of patient records based on reason for referral and ICD codes for PM and DM (1986–1991)	35 with PM 40 with DM	88% Chinese 6.7% Malayan 2.7% Indian 2.7% Other ethnicity Mean age at diagnosis 50.3 years (range 19.2–83.1 years) 65.3% female	Bohan and Peter	0.77	N/A	[27]
Israel	Retrospective review of hospital records for patients with a discharge diagnosis of myositis (1960–1976)	89 with PM or DM	96.6% Jewish, 3.4% Muslim Arab Bimodal age distribution (20.7% <20 years) 59% female	Bohan and Peter	0.22 (increasing with age: 0.05 for 20-29 years vs 0.63 for 60–69 years)	N/A	[28]
Oceania							
South Australia, Australia	Retrospective review of muscle biopsies reported at a central laboratory which reports all samples from the state with subsequent review of associated clinical records (1989–2009)	181 with PM 45 with DM 126 with IBM	Ethnicity not reported Mean age of DM: 55.1 years (range 18–90 years), PM: 59 years (18–86 years), IBM: 67.5 years (range 35–92 years) 59% female (50% for IBM subgroup)	Histopathological criteria	0.8 (0.41 – PM, 0.1 – DM, 0.29 – IBM) Increasing incidence noted from 1980 to 2006 (1.72)	14.2 (7.21 – PM, 1.97 – DM, 5.05 – IBM) 13.9 – IBM when adjusted to >50 years	[29]
South Australia, Australia	Retrospective review of hospital separation summaries (public and private) for ICD-9 codes consistent with	99 with PM 21 with DM	Ethnicity not reported 70-79 age group most common for IIM diagnosis	Histological diagnosis on muscle biopsy ICD codes	0.09 – DM (biopsy-proven) 0.29 – DM (including hospital	N/A	[30]

	PM/DM combined with review of patients with a muscle biopsy showing histological diagnosis of IIM (1991–2005)		57% female		separation ICD codes) 0.44 – PM		
North Canterbury, New Zealand	Retrospective review of hospital clinical records of patients with discharge coding data for IIM with additional case identification from rheumatology and neurology department databases (1989–2001)	17 with PM 18 with DM 6 with IBM 3 with overlap myositis	Ethnicity not reported Mean age at onset 56.6 – PM without malignancy, 60.3 – PM with malignancy, 57.1 – DM without malignancy, 65.2 – DM with malignancy, 72 – IBM, 61.3 – overlap myositis 80% female	Bohan and Peter Mastaglia and Philips for IBM	0.87	N/A	[31]
Victoria, Australia	Retrospective review of all muscle biopsies from the state and review of hospital records with ICD-9 codes to identify IIM (1989–1991)	56 with PM, 12 with DM, 7 with juvenile myositis, 8 with CAM, 11 with overlap myositis	Ethnicity not reported Mean age 55.1 years (SD 19.5 years) 58% female	ICD-9 coding Bohan and Peter Banker and Engel	0.74 No space-time clustering of incidence but spatial clustering was noted	N/A	[32]
Africa							
Assiut Governorate, Upper Egypt	Questionnaire administered door to door for neuromuscular disease with subsequent clinical evaluation and investigations including muscle biopsy if indicated	Various neuromuscular disorders, 6 with IIM (3 with PM, 2 with DM)	Ethnicity not reported Age not reported Sex not reported	Not reported	N/A	11.5 (5.7 – PM, 3.8 – DM)	[33]

	(1996–1997)						
PM							
Águas Formosas, Brazil	Retrospective review of medical records from 26 primary health care units in the region for various autoimmune diseases (2016)	Various autoimmune diseases, one with PM	Ethnicity not reported Age not reported Female patient	Not reported	N/A	1.6	[34]
Kumamoto City, Japan	Survey of medical practitioners requesting patient lists with neuromuscular disease (1977-1982)	Various neuromuscular diseases, 27 with PM	Ethnicity not reported Mean age 44.2 years (range 7–74 years) 81.5% female	Walton–Adams clinical criteria and muscle biopsy	N/A	5	[35]
Benghazi, Libya	Retrospective review of medical records from health-care organisations for patients with neuromuscular disorders (1983–1985)	13 with PM	Ethnicity not reported Mean age 26.6 years (range 3.5–70 years) 69.2% female	DeVere and Bradley	0.84 (1.19 – female, 0.49 – male) Incidence highest in 20–40-year-old group – 1.73 (2.94 – female, 0.56 – male)	N/A	[36]
DM							
Olmsted County, USA	Retrospective population-based cohort study within the Rochester Epidemiology Project to identify incident cases of DM (1995–2019)	29 incident cases of DM (12 – amyopathic, 17 – myopathic)	97% white, non-Hispanic Mean age 57 years (SD 17 years) 90% female	EULAR/ACR 2017 criteria	1.1 (1.9 – female, 0.7 – myopathic DM, 0.5 – CADM) 0.12 (1995–2007) 0.11 (2008–2019) Incidence increased with age	13 (20 – female, 5.8 – myopathic DM, 7.0 – CADM) Prevalence increased with age (34 - >80-years old)	[37]

					(3.2 - >80 years old)		
USA	Retrospective review of people with incident DM in a Veterans' Affairs health care system (2005–2019)	679 with DM	68.2% white, 24.4% black, 4.1% missing Median age 60 years (IQR 52-67 years) 17.5% female	ICD-9 and/or ICD-10 codes	0.8 (1.0 adjusted for age and sex)	N/A	[38]
Olmsted, USA	Retrospective population-based cohort study within the Rochester Epidemiology Project to identify people with DM (1976–2007)	20 with classic DM 3 with ADM 3 with HDM 6 with CADM 3 with initially CADM then myopathic DM 3 with JDM	83% white 3.4% Asian 3.4% Hispanic Mean age 57.4 years (range 30.5–95 years) excluding JDM (mean age 6.8 years) 76% female	Adapted criteria from Gerami et al.	0.96 (1.4 – female, 0.47 – male) adjusted for age 0.21 – ADM, 0.35 – CADM, adjusted for age and sex 0.44 (1976–1985) 1.25 (1996–2007)	21.4 (33.1 – female, 8.4 – males) adjusted for age	[39]
Rural Oaxaca, Mexico	Cross-culturally adapted COPCORD questionnaire performed with people >18 years of the Mixtec and Chontal communities. Subsequent clinical examination and medical record review (time period not reported)	Various rheumatic diseases, one with DM	Mixtec ethnicity Age not reported Sex not reported	Not reported	N/A	100	[40]
Magnesia, Greece	Randomly selected cross-sectional survey (2% of population) followed by clinical examination and	Various rheumatic diseases, one with DM	Not reported	Not reported	N/A	58	[41]

	testing of positive respondents (2007–2008)						
Edirne, Turkey	Retrospective review of patients diagnosed with dermatomyositis at a single centre (Trakya University Medical Faculty) (2004–2014)	23 with DM	Ethnicity not reported Median age 43.6 years (range 17–80 years) 60.9% female	Bohan and Peter	0.37 (0.46 – female, 0.29 – male) 0.45 (2004–2009) 0.29 (2010–2014)	3.2 (3.9 – female, 2.5 – for male)	[42]
South and South-Eastern Botswana	Retrospective review of patient records in Dermatology clinics for autoimmune skin diseases (2008–2015)	Various autoimmune skin diseases, 11 with DM	Ethnicity not reported Age not reported for DM group Gender not reported for DM group	Not reported	0.12	N/A	[43]
sIBM							
Olmsted County, USA	Retrospective population-based cohort study within the Rochester Epidemiology Project to identify patients with sIBM with subsequent manual chart review to apply classification criteria (1980–2019)	21 with sIBM (10 clinicopathologically defined, 9 clinically defined, 1 probable, 1 possible)	100% white Median age at symptom onset 67 years (range 43–86 years) Median age at diagnosis 68 years (range 51–87 years) 47.6% female	ENMC 2011	1.22 (1980–1989), adjusted for age and sex 0.47 (2010–2019), adjusted for age and sex	27.8 (1990, adjusted to >50 years old) 19.2 (2010, adjusted to >50 years old)	[44]
Connecticut, USA	Retrospective review of medical records, EMG and muscle biopsies of patients with a diagnosis of SIBM (1992–2000)	35 with sIBM	Ethnicity not reported Mean age at disease onset 64.3 years (range 41–80 years) 34.3% female	Griggs	N/A	1.1 (0.7 – female, 15 – male) 2.9 (1.8 – female, 4.2 – male) when adjusted for ≥45-years old	[45]

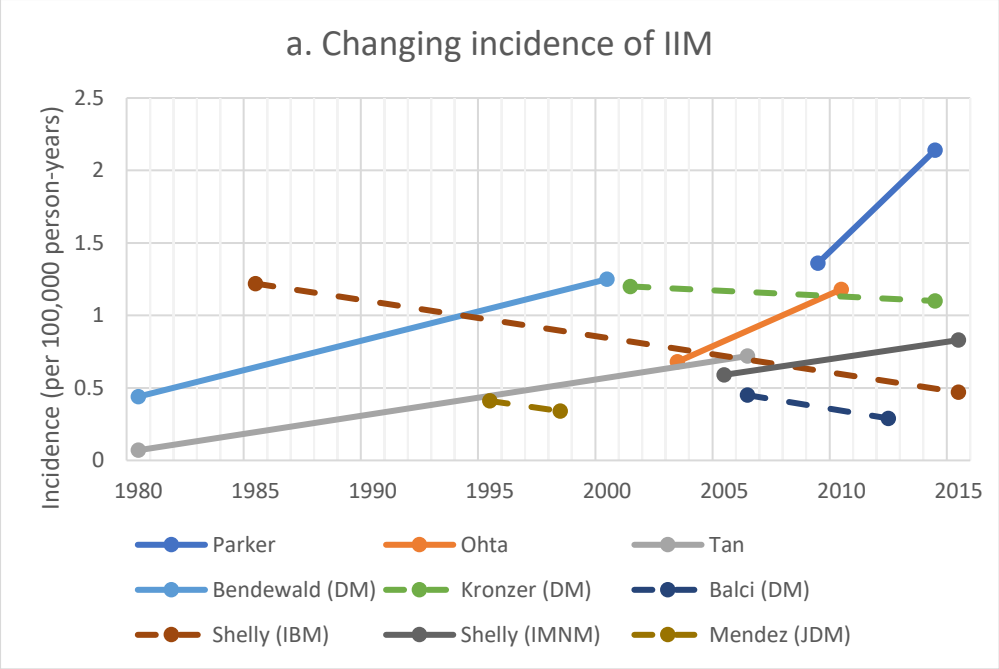
Region Västra Götaland, Sweden	Retrospective review of muscle pathology registry and outpatient clinic registry at Sahlgrenska University Hospital with re-examination of clinical record and muscle biopsy (1985–2017)	128 with clinicopathological criteria sIBM 14 with clinical criteria sIBM	Ethnicity not reported Mean age at onset 66.4 years 30.5% female	ENMC	0.25	3.2 (clinico-pathologically defined) 3.4 (clinically defined)	[46]
Dublin and Cork, Ireland	Prospective and retrospective identification of adults with inherited and acquired neuromuscular disorders, excluding PM/DM (1990–2013)	149 with sIBM	Ethnicity not reported Age not reported Sex not reported	MRC	N/A	11.7	[47]
Southeast Norway	Retrospective review of patient administrative databases for ICD-10 codes with subsequent chart review and muscle biopsy review of these patients to ensure meeting criteria (2003–2012)	95 with sIBM	Ethnicity not reported Mean age at diagnosis 66.9 years (SD 9.3 years) 40% female	ENMC (1997 and/or 2011)	0.2-0.6 annual incidence rate and relatively stable from year to year	3.3 Variation noted around the counties of SE Norway (highest in Telemark – 7)	[48]
Netherlands	Cases identified from 25 specialist neurology or rheumatology centres, computerised coding systems and local pathology databases with subsequent clinical record review and muscle biopsy re-examination	103 with sIBM	100% white Male – mean age 68 years (range 48–84 years); Female – mean age 74 years (range 52–85 years) 34.2% female	ENMC definite or probable SIBM	N/A	0.49 1.6 (1.0 – female, 2.2 – for male) for >50-years old	[49]

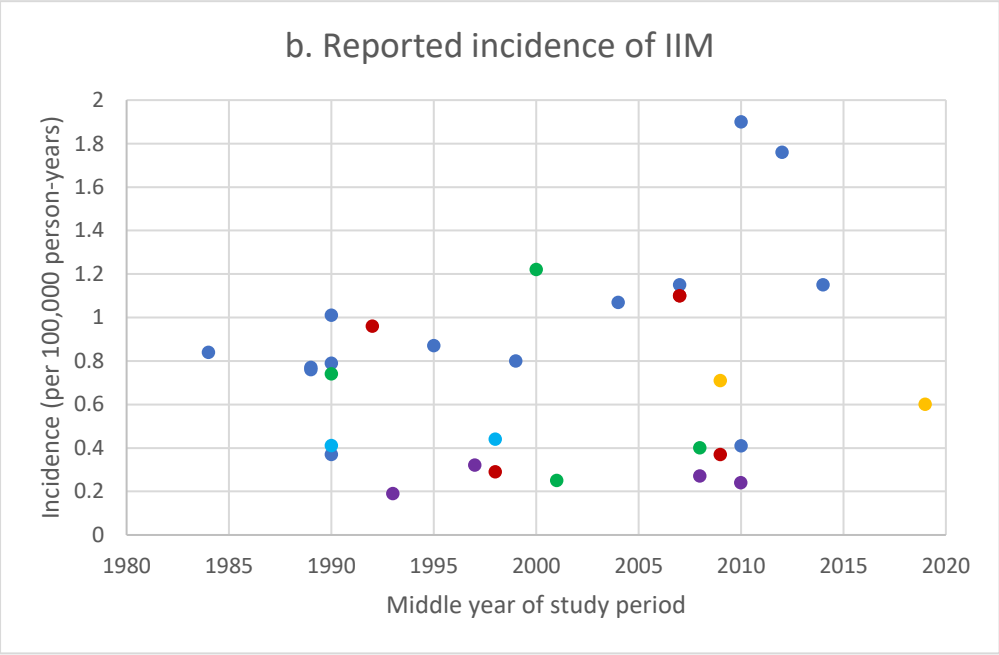
	(1982–1999)						
Istanbul, Turkey	Retrospective re-review of muscle biopsies, using light (but not electron) microscopy, and clinical context on request form to identify cases of sIBM (1993–2011)	9 with biopsy most-suggestive of sIBM 5 with probable sIBM 11 with questionable sIBM	Ethnicity not reported Age not reported Sex not reported	Biopsy (LM but not EM) and request form review	N/A	0.07 (0.38 adjusted for age) when using only most-suggestive sIBM category	[50]
Western Australia, Australia	Retrospective review of all referrals to Australian Neuromuscular Research Institute and muscle biopsy reports. Questionnaire sent to all neurologists and rheumatologists in the state as well to identify patients with IBM (1988–1998)	17 with sIBM (13 definite and 4 possible)	Ethnicity not reported Mean age of onset 49.8 years in females (range 37–56 years) and 60 years in males (range 32–83 years) 41.2% female	Griggs	N/A	0.93 3.53 for >50-years old	[51]
IMNM							
Olmsted County, USA	Retrospective population-based cohort study within the Rochester Epidemiology Project to identify patients with IMNM through ICD codes, specific word searches and laboratory records for anti-HMGCR and anti-SRP seropositivity (1999–2019)	7 with IMNM (4 with anti-HMGCR and statin-exposure, 1 with anti-SRP)	100% white Median age at onset 64 years (range 52–86 years) 28.6% female	ENMC 2016	0.59 (2000–2009) 0.83 (2010–2019)	3.6/1.9 (using 2000/2010 census for denominator, adjusted for age >50 years and sex)	[52]
Cantabria, Spain	Retrospective review of all patients with	10 with IMNM	100% white	ENMC	0.6	3 (all taking statins at the	[53]

	positive anti-HMGCR antibodies (processed at a single immunology laboratory) to identify patients with anti-HMGCR positive IMNM (2016–2021)		Mean age at diagnosis 64.9 years (range 56–79 years) 62.5% female			time of diagnosis)	
JDM							
USA	Retrospective review of the National Institute of Arthritis and Musculoskeletal Skin Disease (NIAMSD) Juvenile Dermatomyositis Research Registry and the national PRDR registry at Indiana University (1994–1999)	395 with JDM	65.1% white, 14.2% Hispanic, 11.4% African American, 1.3% Asian or Pacific Islander, 0.5% American Indian or Alaskan Native Age not reported 69.7% female	Bohan and Peter	0.32 (0.34 – white, 0.33 – African America, 0.27 – Hispanic) 1995 – 0.41 1997 – 0.34	N/A	[54]
Alsace, France	Retrospective multicentre study using a capture-recapture method to identify patients with JDM (2000–2015)	16 with JDM	Ethnicity not recorded Median age 8.6 years (IQR 4–14 years) 75% female	Bohan and Peter or EULAR/ACR 2017	0.27	3.78	[55]
UK	Retrospective review of survey performed of rheumatologists and dermatologists in the country asking how many cases of JDM they had seen in the preceding month (1992–1993)	48 with JDM	88% white 4.2% Black Caribbean 4.2% Pakistani 2.1% Chinese Median age at onset 6.8 years (range 1.3–15.2 years) 83.3% female	Bohan and Peter	0.19	N/A	[56]

Lausanne and Geneva, Switzerland	Retrospective review of patient records at two centres of paediatric rheumatology (1997–2010)	13 with JDM	Ethnicity not reported Mean age 6 years (range 3–10 years) No sex predominance	Clinical evaluation and muscle biopsy, enzymes or imaging	N/A	0.35	[57]
Auckland and surrounding areas, New Zealand	Retrospective review of hospital database using the term "dermatomyositis" for patients <16-years old. (2000–2020)	31 with JDM	51.6% European, 22.6% Maori, 9.7% Pacific Islander, 16.1% Asian Mean age 8.6 years (SD 3.7 years) 58% female	Bohan and Peter	0.24 (no significant difference noted between ethnicities)	N/A	[58]

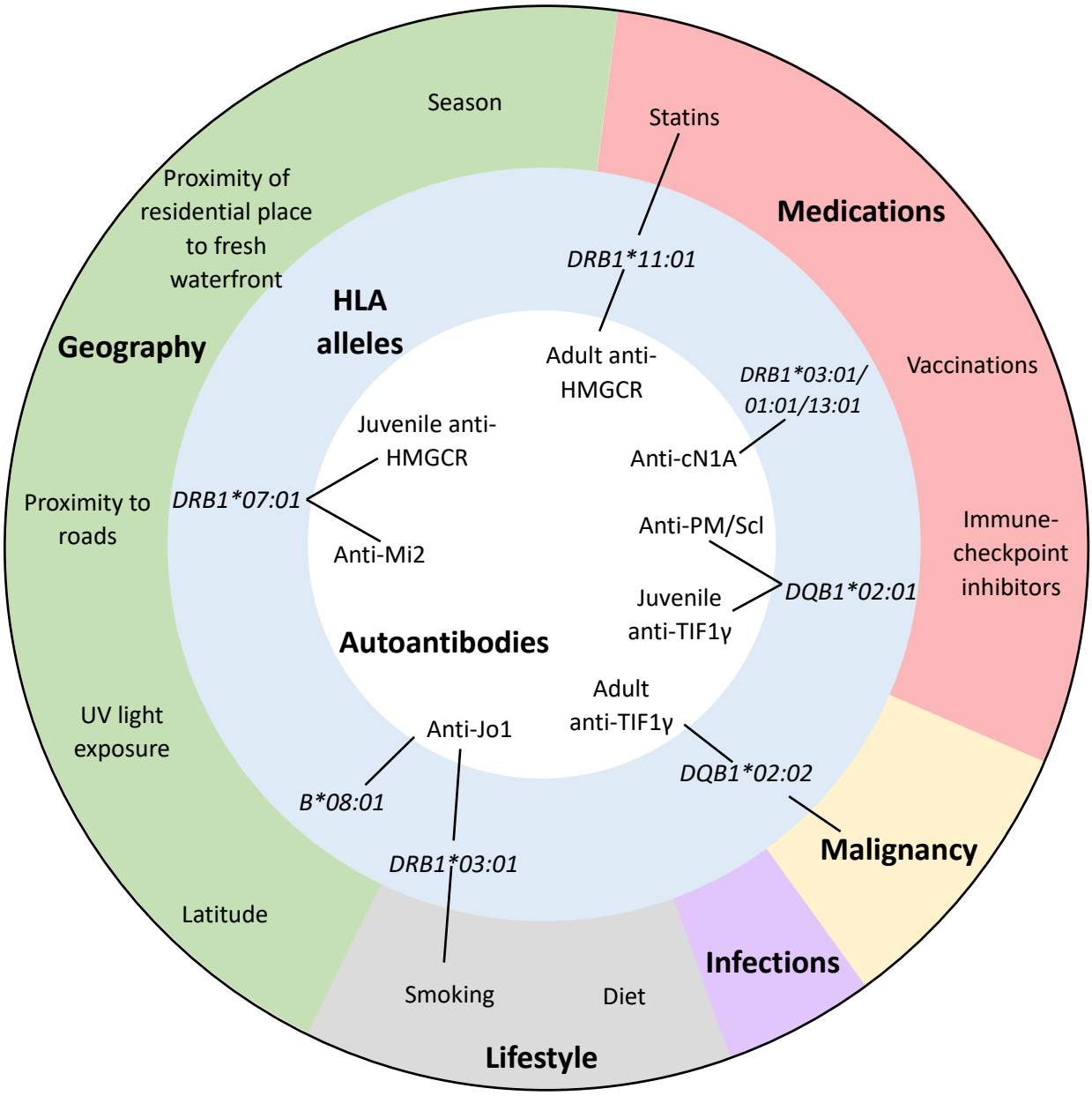
ADM, amyopathic dermatomyositis; CADM, clinically amyopathic dermatomyositis; CAM, cancer-associated myositis; CTD, connective tissue disease; DM, dermatomyositis; EM, electron microscopy; ENMC, European Neuromuscular Centre; HDM, hypomyopathic dermatomyositis; IMNM, immune-mediated necrotizing myopathy; JDM, juvenile dermatomyositis; LM, light microscopy; MRC, medical research council; sIBM, sporadic inclusion-body myositis.





Supplementary Figure 2 | Longitudinal and average incidences reported in epidemiological studies of idiopathic inflammatory myopathies.

a) Changing incidence of IIM where reported at different timepoints within studies that utilized a form of case verification beyond ICD disease coding alone (solid line – increasing incidence, dashed line – decreasing incidence). Refs: Parker[13], Bendewald[39], Shelly (IBM)[44], Ohta[22], Kronzer[37], Shelly (IMNM)[52], Tan[29], Balci[42], Mendez[54]. b) Average incidence of IIM reported in epidemiological studies: mixed IIM subtypes (blue), DM (red), PM (teal), sIBM (green), IMNM (yellow), JDM (purple). DM, dermatomyositis; IIM, idiopathic inflammatory myopathies; IMNM, immune-mediated necrotizing myopathy; JDM, juvenile dermatomyositis; PM, polymyositis; sIBM, sporadic inclusion body myositis.



Supplementary Figure 3 | Interaction of environmental and genetic risk factors with autoantibodies in white patients with idiopathic inflammatory myopathies.

References:

1 Furst, D. E., Amato, A. A., Iorga, S. R., Gajria, K. & Fernandes, A. W. Epidemiology of adult idiopathic inflammatory myopathies in a U.S. managed care plan. *Muscle and Nerve* **45**, 676-683 (2012).

2 Smoyer-Tomic, K. E., Amato, A. A. & Fernandes, A. W. Incidence and prevalence of idiopathic inflammatory myopathies among commercially insured, Medicare supplemental insured, and Medicaid enrolled populations: An administrative claims analysis. *BMC Musculoskeletal Disorders* **13**, 103 (2012).

3 Barnabe, C. *et al.* Prevalence of autoimmune inflammatory myopathy in the first nations population of Alberta, Canada. *Arthritis Care and Research* **64**, 1715-1719 (2012).

4 Bernatsky, S. *et al.* Estimating the prevalence of polymyositis and dermatomyositis from administrative data: age, sex and regional differences. *Ann Rheum Dis* **68**, 1192-1196 (2009).

5 Wilson, F. C., Ytterberg, S. R., St. Sauver, J. L. & Reed, A. M. Epidemiology of sporadic inclusion body myositis and polymyositis in Olmsted County, Minnesota. *Journal of Rheumatology* **35**, 445-447 (2008).

6 Medsger, T. A., Jr., Dawson, W. N., Jr. & Masi, A. T. The epidemiology of polymyositis. *Am J Med* **48**, 715-723 (1970).

7 Bernal-Macias, S., Fernandez-Avila, D. G., Rincon-Riano, D. N., Gutierrez, J. M. & Rosselli, D. Prevalence of inflammatory myopathies in Colombia: Demographic analysis from the National Health Registry 2012-2018. *Revista Colombiana de Reumatologia* (2021).

8 Rosa, J. *et al.* Incidence and prevalence of polymyositis and dermatomyositis in a health management organization in Buenos Aires. *Journal of Clinical Rheumatology* **19**, 303-307 (2013).

9 Pawlitzki, M. *et al.* Myositis in Germany: epidemiological insights over 15 years from 2005 to 2019. *Neurological Research and Practice* **4**, 62 (2022).

10 Carey, I. M. *et al.* Prevalence and incidence of neuromuscular conditions in the UK between 2000 and 2019: A retrospective study using primary care data. *PLoS ONE* **16**, e0261983 (2021).

11 Siso-Almirall, A. *et al.* The prevalence of 78 autoimmune diseases in Catalonia (MASCAT-PADRIS Big Data Project). *Autoimmunity Reviews* **19**, 102448 (2020).

12 Ostrovršnik, J. *et al.* The incidence of idiopathic inflammatory myopathies in the adult Slovenian population. *Clin Rheumatol* **38**, 279-283 (2019).

13 Parker, M. J. S., Oldroyd, A., Roberts, M. E., Cooper, R. G. & Chinoy, H. Increasing incidence of adult idiopathic inflammatory myopathies: A ten-year UK epidemiological study. *Annals of the Rheumatic Diseases* **77**, 757 (2018).

14 Svensson, J., Arkema, E. V., Lundberg, I. E. & Holmqvist, M. Incidence and prevalence of idiopathic inflammatory myopathies in Sweden: A nationwide population-based study. *Rheumatology (United Kingdom)* **56**, 802-810 (2017).

15 Elfving, P. *et al.* Estimating the incidence of connective tissue diseases and vasculitides in a defined population in Northern Savo area in 2010. *Rheumatology International* **36**, 917-924 (2016).

16 Dobloug, C. *et al.* Prevalence and clinical characteristics of adult polymyositis and dermatomyositis; Data from a large and unselected Norwegian cohort. *Annals of the Rheumatic Diseases* **74**, 1551-1556 (2015).

17 Eaton, W. W., Rose, N. R., Kalaydjian, A., Pedersen, M. G. & Mortensen, P. B. Epidemiology of autoimmune diseases in Denmark. *Journal of Autoimmunity* **29**, 1-9 (2007).

18 Weitoft, T. Occurrence of polymyositis in the county of Gavleborg, Sweden. *Scandinavian Journal of Rheumatology* **26**, 104-106 (1997).

19 Kaipiainen-Seppanen, O. & Aho, K. Incidence of rare systemic rheumatic and connective tissue diseases in Finland. *Journal of Internal Medicine* **240**, 81-84 (1996).

20 Ahlstrom, G., Gunnarsson, L. G., Leissner, P. & Sjoden, P. O. Epidemiology of neuromuscular diseases, including the postpolio sequelae, in a Swedish county. *Neuroepidemiology* **12**, 262-269 (1993).
21 Cho, S. K. *et al.* Incidence and Prevalence of Idiopathic Inflammatory Myopathies in Korea: a Nationwide Population-based Study. *Journal of Korean medical science* **34**, e55 (2019).
22 Ohta, A., Nagai, M., Nishina, M., Tomimitsu, H. & Kohsaka, H. Prevalence and incidence of polymyositis and dermatomyositis in Japan. *Modern Rheumatology* **24**, 477-480 (2014).
23 See, L. C., Kuo, C. F., Chou, I. J., Chiou, M. J. & Yu, K. H. Sex- and age-specific incidence of autoimmune rheumatic diseases in the Chinese population: A Taiwan population-based study. *Seminars in Arthritis and Rheumatism* **43**, 381-386 (2013).
24 Yu, K. H., See, L. C., Kuo, C. F., Chou, I. J. & Chou, M. J. Prevalence and incidence in patients with autoimmune rheumatic diseases: A nationwide population-based study in Taiwan. *Arthritis Care and Research* **65**, 244-250 (2013).
25 Kuo, C. F. *et al.* Incidence, cancer risk and mortality of dermatomyositis and polymyositis in Taiwan: A nationwide population study. *British Journal of Dermatology* **165**, 1273-1279 (2011).
26 Kusumi, M., Nakashima, K., Nakayama, H. & Takahashi, K. Epidemiology of inflammatory neurological and inflammatory neuromuscular diseases in Tottori Prefecture, Japan. *Psychiatry and Clinical Neurosciences* **49**, 169-174 (1995).
27 Koh, E. T. *et al.* Adult onset polymyositis/dermatomyositis: clinical and laboratory features and treatment response in 75 patients. *Ann Rheum Dis* **52**, 857-861 (1993).
28 Benbassat, J., Geffel, D. & Zlotnick, A. Epidemiology of polymyositis-dermatomyositis in Israel, 1960-76. *Israel Journal of Medical Sciences* **16**, 197-200 (1980).
29 Tan, J. A. *et al.* Incidence and prevalence of idiopathic inflammatory myopathies in South Australia: a 30-year epidemiologic study of histology-proven cases. *International journal of rheumatic diseases* **16**, 331-338 (2013).
30 Limaye, V. *et al.* The epidemiology of dermatomyositis in South Australia. *APLAR Journal of Rheumatology* **10**, 94-100 (2007).
31 Lynn, S. J., Sawyers, S. M., Moller, P. W., O'Donnell, J. L. & Chapman, P. T. Adult-onset inflammatory myopathy: North Canterbury experience 1989-2001. *Internal Medicine Journal* **35**, 170-173 (2005).
32 Patrick, M., Buchbinder, R., Jolley, D., Dennett, X. & Buchanan, R. Incidence of inflammatory myopathies in Victoria, Australia, and evidence of spatial clustering. *Journal of Rheumatology* **26**, 1094-1100 (1999).
33 El-Tallawy, H. N., Khedr, E. M., Qayed, M. H., Helliwell, T. R. & Kamel, N. F. Epidemiological study of muscular disorders in Assiut, Egypt. *Neuroepidemiology* **25**, 205-211 (2005).
34 Tolentino, D. S., de Oliveira, C. M. & de Assis, E. M. Population-based study of 24 autoimmune diseases carried out in a brazilian microregion. *Journal of Epidemiology and Global Health* **9**, 243-251 (2019).
35 Araki, S., Uchino, M. & Kumamoto, T. Prevalence studies of multiple sclerosis, myasthenia gravis, and myopathies in Kumamoto district, Japan. *Neuroepidemiology* **6**, 120-129 (1987).
36 Radhakrishnan, K., El-Mangoush, M. A. & Gerryo, S. E. Descriptive epidemiology of selected neuromuscular disorders in Benghazi, Libya. *Acta Neurologica Scandinavica* **75**, 95-100 (1987).
37 Kronzer, V. L. *et al.* Incidence, Prevalence, and Mortality of Dermatomyositis: A Population-Based Cohort Study. *Arthritis Care and Research* **75**, 348-355 (2023).
38 Bolender, C. M. *et al.* Incidence of Dermatomyositis in a Nationwide Cohort Study of US Veterans. *JAMA Dermatology* **158**, 1321-1323 (2022).
39 Bendewald, M. J., Wetter, D. A., Li, X. & Davis, M. D. Incidence of dermatomyositis and clinically amyopathic dermatomyositis: a population-based study in Olmsted County, Minnesota. *Arch Dermatol* **146**, 26-30 (2010).

40 Julian-Santiago, F., Garcia-Garcia, C., Garcia-Olivera, I., Goycochea-Robles, M. V. & Pelaez-Ballestas, I. Epidemiology of rheumatic diseases in Mixtec and Chontal indigenous communities in Mexico: a cross-sectional community-based study. *Clinical Rheumatology* **35**, 35-42 (2016).
41 Anagnostopoulos, I. *et al.* The prevalence of rheumatic diseases in central Greece: A population survey. *BMC Musculoskeletal Disorders* **11**, 98 (2010).
42 Balci, M. A., Donmez, S., Saritas, F., Bas, V. & Pamuk, O. N. The epidemiology of dermatomyositis in northwestern Thrace region in Turkey: epidemiology of dermatomyositis in Turkey. *Rheumatology International* **37**, 1519-1525 (2017).
43 Madu, P. N. *et al.* Autoimmune skin disease among dermatology outpatients in Botswana: a retrospective review. *Int J Dermatol* **58**, 50-53 (2019).
44 Shelly, S. *et al.* Epidemiology and Natural History of Inclusion Body Myositis: A 40-Year Population-Based Study. *Neurology* **96**, e2653-e2661 (2021).
45 Felice, K. J. & North, W. A. Inclusion body myositis in Connecticut: Observations in 35 patients during an 8-year period. *Medicine* **80**, 320-327 (2001).
46 Lindgren, U., Pullerits, R., Lindberg, C. & Oldfors, A. Epidemiology, Survival, and Clinical Characteristics of Inclusion Body Myositis. *Annals of Neurology* **92**, 201-212 (2022).
47 Lefter, S., Hardiman, O. & Ryan, A. M. A population-based epidemiologic study of adult neuromuscular disease in the Republic of Ireland. *Neurology* **88**, 304-313 (2017).
48 Dobloug, G. C. *et al.* High prevalence of inclusion body myositis in Norway; a population-based clinical epidemiology study. *European Journal of Neurology* **22**, 672-e641 (2015).
49 Badrising, U. A. *et al.* Epidemiology of inclusion body myositis in the Netherlands: A nationwide study. *Neurology* **55**, 1385-1387 (2000).
50 Serdaroglu Oflazer, P., Deymeer, F. & Parman, Y. Sporadic-inclusion body myositis (s-IBM) is not so prevalent in Istanbul/Turkey: A muscle biopsy based survey. *Acta Myologica* **30**, 34-36 (2011).
51 Phillips, B. A., Zilko, P. J. & Mastaglia, F. L. Prevalence of sporadic inclusion body myositis in Western Australia. *Muscle and Nerve* **23**, 970-972 (2000).
52 Shelly, S. *et al.* Incidence and prevalence of immune-mediated necrotizing myopathy in adults in Olmsted County, Minnesota. *Muscle and Nerve* **65**, 541-546 (2022).
53 Prieto-Pena, D. *et al.* Epidemiological and genetic features of anti-3-hydroxy-3-methylglutaryl-CoA reductase necrotizing myopathy: Single-center experience and literature review. *Eur J Intern Med* **101**, 86-92 (2022).
54 Mendez, E. P. *et al.* US incidence of juvenile dermatomyositis, 1995-1998: Results from the National Institute of Arthritis and Musculoskeletal and Skin Diseases Registry. *Arthritis Care and Research* **49**, 300-305 (2003).
55 Moegle, C., Severac, F. & Lipsker, D. Epidemiology of juvenile dermatomyositis in Alsace. *British Journal of Dermatology* **182**, 1307-1308 (2020).
56 Symmons, D. P. M., Sills, J. A. & Davis, S. M. The incidence of juvenile dermatomyositis: Results from a nation-wide study. *British Journal of Rheumatology* **34**, 732-736 (1995).
57 Bellutti Enders, F. & Hofer, M. Juvenile dermatomyositis in Western Switzerland. *Pediatric Rheumatology* **9** (2011).
58 Concannon, A. & Han, D. Y. Incidence, severity and clinical manifestations of juvenile dermatomyositis among Maori and Pacific Island compared to European children. *Journal of Paediatrics and Child Health* **57**, 1881-1885 (2021).