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Review Article **Open Access**

Ethnicity association in primary systemic vasculitis: A systematic search and review

Moshiur Rahman Khasru[1]*, Tangila Marzen[2], Mohammad Tariqul Islam[1], Khandakar Mohammad Hossain[3], Badrunnesa Ahmed[1], Abul Khair Mohammad Salek[1], Md. Sharfuddin Ahmed[4], Mohamed Sakel[5], Nasrin Akter[6] Denny Clegg[7]

[1] Department of Physical Medicine and Rehabilitation, Bangabandhu Sheikh Mujib Medical University, Dhaka-1000, Bangladesh. [2] Department of Dental Radiology (Deputed in Anatomy Department), Shaheed Suhrawardy Medical College, Dhaka- 1207, Bangladesh. [3] Department of Paediatrics, Combined Military Hospital, Dhaka Cantonment, Dhaka, Bangladesh. [4] Office of the Vice Chancellor, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh. [5] Department of Neuro Rehabilitation, East Kent Hospital University, Canterbury, United Kingdom. [6] Department of Pharmacology, Care Medical College, Dhaka, Bangladesh. [7] Department of Rheumatology, University of South Wales, Cardiff, United Kingdom.

Literature described wide disparities in incidence and prevalence between different types of vasculitis. There were no comprehensive studies on ethnic or racial associations in all types of primary systemic vasculitis (PSV) in any published article until this review commenced in 2020. The review aims to synthesize the evidence regarding the relation between ethnicity and the incidence and/or prevalence of different types of PSV. A total of 52 selected articles which include clinical trials, cohorts, cross-sectional studies, case series, and case studies and have been published within the last 10 years in the human population, were reviewed by searching Medline, PubMed, and Google Scholars databases using predefined keywords. The PRISMA diagrams were followed to identify relevant articles. The methodological qualities of the studies were assessed using the EPHPP tool. Finally, a summary of the evidence on the association between ethnic origin and PSV was painstakingly compiled. The connection between ethnicity and different types of PSV has been found to be significantly diverse in this researchas vasculitis is more common in Asians and Scandinavians, Kawasaki disease and periarteritis nodules are more prevalent in Japanese and Alaskanatives,ANCAassociated vasculitis is more frequent in Caucasians, whereas Henoch-Schonleinpurpura and Cogan syndrome more usual in Caucasians and Asians. Furthermore, Behçet's disease more commonly occurs on the "Silk Road," especially in Turkey. Genetic susceptibility and environmental elements could contribute to the global variation in the incidence and prevalence of primary systemic vasculitis.

Keywords: Vasculitis; Ethnic; Racial;Association; Incidence; Prevalence.

Introduction

Vasculitis is a rare heterogeneous group of disorders that may occur independently, such as (i) primary systemic vasculitis (PSV), e.g., granulomatosis with polyangiitis (GPA) or (ii) secondary to other diseases such as systemic

*Corresponding author: Moshiur Rahman Khasru, Associate Professor and Coordinator, Rheumatology Rehab Clinic Musculoskeletal Medicine and Interventional Physiatrist Division, Department of Physical Medicine and Rehabilitation Bangabandhu Sheikh Mujib Medical University Dhaka-1000, Bangladesh. E-mail: drkhasrupmr@bsmmi.edu.bd

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lupus erythematosus and rheumatoid arthritis. The word "vasculitis" means inflammation of the blood vessels [1]. Vasculitis is usually a systemic disease, although it may be localized to a single organ or vascular bed and sometimes have no clinical significance [1]. The pathological consequence of inflammation observed in histopathology is vascular wall destruction and fibrinoid necrosis [1]. Although the cause of vasculitis is unknown, several factors such as ethnicity, genetic affiliation, sex, and environment, including infections, toxins, ultraviolet rays, medications, smoking, and substances,have been identified cause allergies to influence the manifestation of the disease [1]. As terms "vasculitis", and "arteritis", include about 20 different types of vasculitis [2]. Vasculitides are classified according to vessel size and the serum's antineutrophil cytoplasmic antibodies (ANCA). However, the nomenclature of Chapel Hill Consensus 2012 (CHCC2012) is generally considered for the classification of vasculitis [3]. According to CHCC2012, Takayasu's arteritis (TAK) and giant cell arteritis (GCA) belongs to the group of large vessel inflammatory diseases. Moderate vasculitis includes periarterial nodal inflammation (PAN) and Kawasaki disease (KD). Small-vessel-vasculitis are (a) ANCA associated vasculitis (AAV) including (i) microscopic polyangiitis (MPA), (ii) granulomatosis with polyangiitis (GPA), and (iii) eosinophilic granulomatosis with polyangiitis (EGPA), and (b) immunocomplex vasculitis including (i) antiglomerular basement membrane disease (AntiGBM), (ii) cryoglobulinemia (CV) vasculitis, (iii) Henoch-Schoenleinpurpura (HSP) and (iv) complement deficiency urticarial vasculitis (HUV). In addition, the European Medicines Agency (EMA) vasculitis classification algorithm was recently developed to reconcile the differences between the ACR and CHCC classifications for GPA, MPA, EGPA, and PAN [1].

The incidence of vasculitis, especially ANCA induced vasculitis, has increased significantly over the past 10 years for a variety of reasons, including better knowledge of these diseases. Still, it remains rare, with fewer than 30 new cases per million population yearly [1].

Sufficient evidence establishes that giant cell arteritis (GCA) is a heterogeneous disorder of the elderly regarding clinical features such as fatigue, weight loss, low-grade fever, immunopathology, and its response to treatment [4]. The estimated incidence of GCA varies from 1.4 to 20 per 100,000 people over 50 years of age [5]. According to Crow et al. (2009), the median survival time for GCA is estimated to be 3.7 years [6]. Takayasu's arteritis (TA) is the most common childhood vasculitis and the most common cause of renal vascular hypertension in children. The 5 and 10-year TArelated mortality rates have been estimated at 1.9 and 3.9 per 100 cases, respectively, with higher mortality rates among Caucasians and smokers [7]. The annual incidence of Takayasu's arteritis is about two per million of the population [8]. The prevalence of periarteritis nodules has decreased over the past decades, as has the rate of hepatitis B virus (HBV) infection, but cutaneous periarteritis remains fairly common [1]. Kawasaki disease (KD) is the most common moderate inflammatory acute vasculitis in infants and young children that may get complicated by coronary aneurysms in 1 in 4 untreated cases [9]. Cases of KD have been described as the leading cause of acquired heart disease worldwide in children of developed countries. The prevalence of Kawasaki disease varies widely between 5 and 265 per 100,000 children under 5 years of age [10]. The annual incidence of periarteritis nodule (PAN) varies from 8 to 31 cases per million [11]. The overall incidence of ANCA-associated vasculitis (AVV) ranges from 46 to 184 per million [12]. Among AAVs, the annual incidence of MPA, GPA, and EGPA ranged from 1 to 8; 10 to 95; and <1 to 14 per million, respectively (Hoang and Park, 2020). The annual incidence of HSP ranges from 13 to 127 per million people [11].

The risk factors for vasculitis are also diverse [11]. Demirkaya and colleagues found ethnicity, genetic predisposition, age, and environmental factors common risk factors for PSV [13, 14]. Among people of European ancestry, the HLA DP6-inherited population was approximately 4 times more likely to develop vasculitis than the control population. At the same time, SEPINA1*14 gene-positive individuals showed

an altered 50% lower change in the development of vasculitis compared with the control population [14]. According to Brogan and Eleftheriou (2017), the detection of biomarkers such as proteinase-3 myeloperoxidase (MPO), circulating microparticles, neutrophil extra-cellular trap (NET), and CD163 in urine promote the growing understanding of the pathogenesis of PSV and its prognosis [15]. PAN is associated with the absence of adenosine deaminase type 2 (DADA2) [15]. The clinical features of vasculitis have been shown to vary between different ethnic groups and geographical regions [16, 17]. For example, MPO and anticytoplasmic antibody (ANCA) incidence is higher in Asian countries. On the other hand, Northern Europe and the United States (US) have a higher incidence of proteinase-3 (PR3) ANCA-associated granulomatosis with polyangiitis (GPA) [17]. Pearce and colleagues found large differences in the incidence of ANCA-associated vasculitis (AAV) among eight different ethnic groups in their multicenter observational study, including 1217 patients with AAV at 133 locations from September 2014 to March 2016 [18]. The incidence and prevalence of AAV have increased over the past decade [19]. In recent years, environmental and genetic factors have contributed largely to the increased risk of PSV [19].

To the authors' knowledge, and after extensive research, no comprehensive studies regarding the association between ethnicity or race and the incidence and/or prevalence of primary systemic vasculitis (PSV) are available in a single article before starting this review. Synoptic information regarding ethnic influence in primary vasculitis has the potential to greatly assist epidemiologists, clinical researchers, and clinicians in their practice. This review was planned to answer the questions related to the evidence of the relation between ethnicity and PSV. The review's objectives were (i) to synthesize evidence regarding an ethnic association in different types of PSVand (ii) to determine the incidence and/or prevalence of the different types of PSV. The authors present clinical trials, cohort studies, cross sectional studies, case series, and case studies. This review appears to be an appropriate approach to evidence dissemination, with the primary goal of more clearly answering the question of prevalence and ethnicity in PSV. Search strategy to identify studies: Available evidence was systematically searched in Medline, PubMed, and Google Scholars using the keywords 'vasculitis', 'vasculitis', 'risk factors', 'ethnicity', population race, 'race' and 'racial association', as well as Boolean expressions where appropriate. Articles published in English literature within the past 10 years were considered for this review, and all other articles were automatically excluded. The search was completed on October 31, 2020.

Method and Materials

PRISMA diagrams were followed to identify relevant articles. 182 articles were found in English literature by searching through a search engine using predefined keywords (shown in the PRISMA diagram, Figure 1). Finally, 52 articles [18, 20-70] were selected for this review obeying the selection criteria (shown in Table 1). Then, the methodological quality of the selected articles was assessed using the Effective Public Health Practice Project (EPHPP) – a quality assessment tool for quantitative studies in this review shown in Table 2. Thus, the frequency, incidence, and prevalence of PSV among different ethnic/racial groups were extracted from selected articles according to the selection criteria. All extracted data were then summarized in Table1, showing the author's name, year of publication, country, and reported types of vasculitis. Effect sizes and results are reported in articles and saved in spreadsheets in Microsoft Excel 2016. Extracted data were aggregated and analyzed using the same Microsoft Excel 2016. Finally, a synopsis of evidence on the association between ethnicity/ racial origin and PSV was made meticulously.

Results

This review included 52 publications with 46616166 participants (Table 1). A cross sectional design was used in 46.2% of the articles as the most common study design (24 out of 52). Case-control designs were used in 32.7% of research, followed by cohort studies

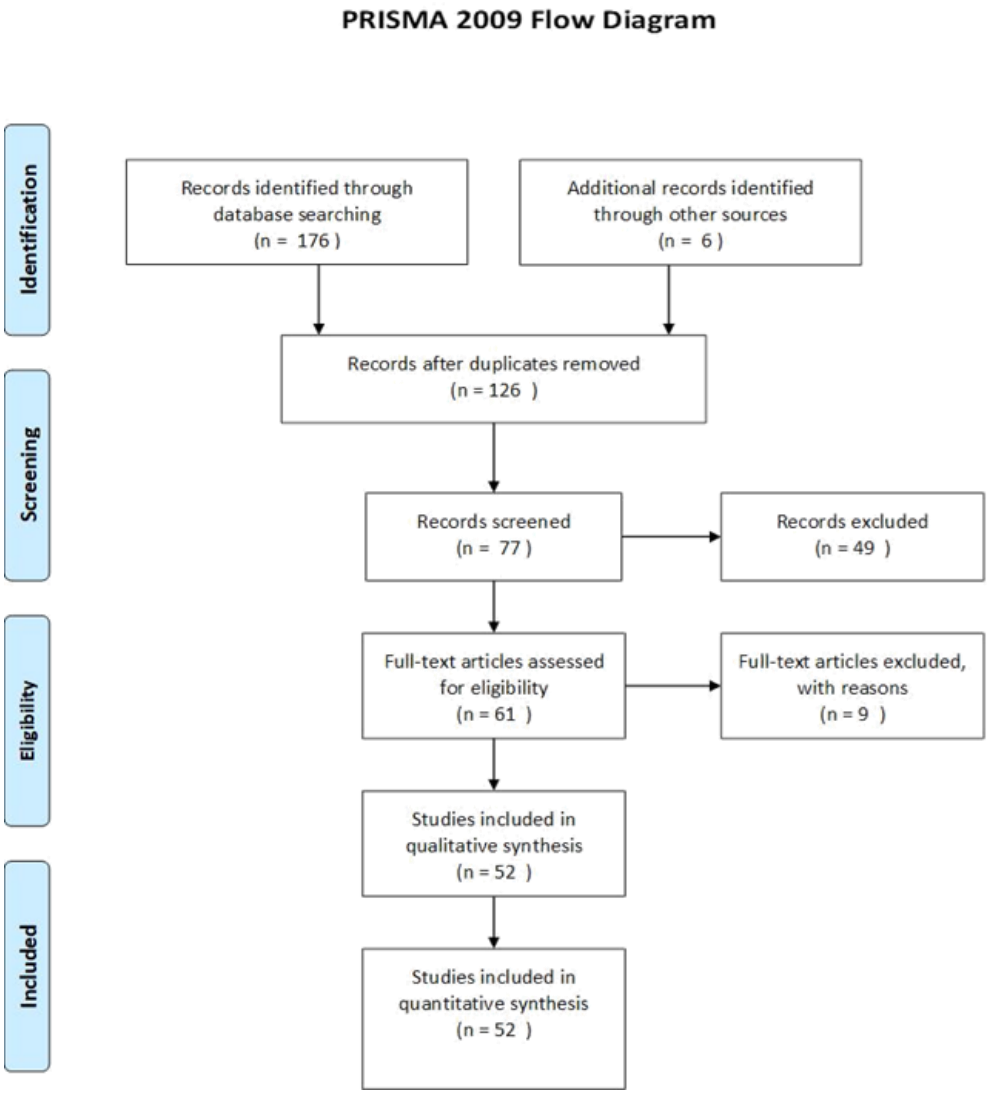

Figure 1. Prisma flow diagram

in 17.3% of cases and case series designs in the remaining studies. The USA, Japan, and China contributed 19.2%, 17.3%, and 17.3% of the articles. Among all enlisted articles, the most frequent articles were around Kawasaki disease 15 (28.9%), taken after by ANCA-related vasculitis 13 (25%), and Behcet's illness 12 (23.1%) individually. The conveyance of articles agreeing to distinctive consideration plans concerning different sorts of vasculitis was displayed within the Figure 2. The affiliation of ethnicity and distinctive types of PSV widely aries, appeared in Table 3. Takayasu disease and giant cell arteritis are more predominant among Asians and Scandinavians. The prevalence of Kawasaki disease and Polyarteritis nodosa are more among people of the Japanese and native Alaskan populace. ANCA related vasculitis is more common among Caucasians in Minnesota, USA. Henoch schonlein purpura and Cogan's syndrome occurs more among Caucasians and Asians. Other than that, Behcet's disease is more predominant within the 'Silk-road', particularly in Turkey. On the other hand, Takayasu disease and giant cell arteritis are less frequent among Jews and Japanese, respectively. Kawasaki disease and polyarteritisnodosa are less common among native Australians, European children, and south Sweden. ANCA-associated vasculitis is less prevalent among black sub-Saharans. Behcet's disease is less frequent among native British.

Table 1. Effective Public Health Practice Project (EPHPP) - Quality assessment tool for quantitative studies

Particulars of the Studies					EPHPP Components						
Sl No.	Author	Year	**Journal**	**Study Design**	**Selection Bias**	**Study design**	**Confounders**	**Blinding**	**Data Collection Method**	**Withdrawal and Dropout**	**Global Rating**
1.	Nair et al. [20]	2018	International Journal of Rheumatic Diseases	Case Control study	M	W	M	NA	S	NA	Moderate
2.	Takamura et al. [21]	2012	Circulation Journal	Case Control study	M	W	S	NA	S	NA	Moderate
3.	Terao et al. [22]	2013	The American Journal of Human Genetics	Cohort study	S	W	NA	NA	M	M	Moderate
4.	Gudbrandsson et al. [23]	2016	Arthritis Care & Research	Cohort study	S	W	S	NA	S	S	Moderate
5.	Bilge et al. [24]	2012	Rheumatology International	Cross Sectional study	W	W	NA	NA	M	NA	Weak
6.	Gruener et al. [25]	2019	JAMA Ophthalmology	Cohort study	S	W	NA	NA	S	S	Moderate
7.	Brekke et al. [26]	2017	Arthritis Research & Therapy	Cohort study	M	W	NA	NA	M	M	Moderate
8.	Tan et al. [27]	2019	Eye	Cross Sectional study	W	W	NA	NA	S	NA	Weak
9.	Pereira et al. [28]	2015	British Journal of Ophthalmology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
10.	Wang et al. [29]	2019	Bioscience Reports	Case control study	W	W	NA	W	S	M	Weak
11.	Kuo et al. [30]	2011	Journal of Human Genetics	Case control study	W	W	W	W	S	M	Weak
12.	Yeter et al. [31]	2016	International Journal of Environmental Research and Public Health	Case control study	W	W	M	W	S	M	Weak
13.	Kim et al. [32]	2017	PLoS ONE	Case series	W	W	W	W	S	S	Weak
14.	Amano et al. [33]	2019	Pediatric Rheumatology	Clinical trial	W	M	M	W	S	S	Weak
15.	Nagelkerke et al. [34]	2019	Frontiers in Immunology	Cohort study	M	W	S	NA	M	M	Moderate
16.	Boudiaf et al. [35]	2016	Journal of Tropical Pediatrics	Cross Sectional study	W	W	NA	NA	S	NA	Weak
17.	Fernandez-Cooke et al. [36]	2019	PLoS ONE	Cross Sectional study	W	W	NA	NA	S	NA	Weak
18.	Holman et al. [37]	2011	Hawaii Medical Journal	Cross Sectional study	W	W	NA	NA	S	NA	Weak
19.	Ghimire et al. [38]	2019	Cardiology in the Young	Cross Sectional study	W	W	NA	NA	S	NA	Weak
20.	Manlhiot et al. [39]	2017	Canadian Journal of Cardiology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
21.	Nakamura et al. [40]	2011	Journal of Epidemiology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
22.	Makino et al. [41]	2015	Journal of Epidemiology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
23.	Makino et al. [42]	2018	Pediatrics International	Cross Sectional study	W	W	NA	NA	S	NA	Weak
24.	Park et al. [43]	2011	Pediatrics International	Cross Sectional study	W	W	NA	NA	S	NA	Weak
25.	Karadag et al. [44]	2018	Rheumatology International	Cohort study	M	W	M	NA	M	M	Moderate
26.	Jelusic et al. [45]	2012	Rheumatology International	Cross sectional study	W	W	NA	NA	S	NA	Weak
27.	Pearce et al. [18]	2017	Rheumatology	Cross sectional study	W	W	NA	NA	S	NA	Weak
28.	Berti et al. [46]	2017	Arthritis & Rheumatology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
29.	Wu et al. [47]	2015	International Journal of Rheumatic Diseases	Case Control study	W	W	S	W	S	M	Weak
30.	Fujimoto et al. [48]	2011	Rheumatology	Case control study	W	W	M	W	S	M	Weak
31.	Cao et al. [49]	2011	Journal of the American Society of Nephrology	Case control study	W	W	S	W	S	M	Weak
32.	Enkhmaa et al. [50]	2015	Atherosclerosis	Case control study	W	W	S	W	S	M	Weak
33.	Ahn et al. [51]	2012	Rheumatology International	Cohort study	M	W	M	NA	M	M	Moderate
34.	Li et al. [52]	2018	Arthritis Research & Therapy	Cross Sectional study	W	W	NA	NA	S	NA	Weak
35.	Faurschou et al. [53]	2013	Clinical and Experimental Rheumatology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
36.	Terrier et al. [54]	2017	Rheumatology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
37.	Nesher et al. [55]	2016	The Journal of Rheumatology	Cross Sectional study	W	W	NA	NA	S	NA	Weak

38.	Wada et al. [56]	2012	The Journal of Rheumatology	Cross Sectional study	W	W	NA	NA	S	NA	Weak
39.	Pearce et al. [57]	2016	Rheumatology	Cross sectional study	W	W	NA	NA	S	NA	Weak
40.	Ortiz-Fernadez et al. [58]	2016	Clinical and Experimental Rheumatology	Cohort study	M	W	M	NA	M	M	Moderate
41.	Saleh et al. [59]	2012	Ocular Immunology and Inflammation	Cross Sectional study	W	W	NA	NA	S	NA	Weak
42.	Li et al. [60]	2012	Human Genetics	Case Control study	W	W	M	NA	S	NA	Weak
43.	Hou et al. [61]	2013	Human Genetics	Case Control study	W	W	S	NA	S	NA	Weak
44.	Wu et al. [62]	2014	Clinical and Experimental Rheumatology	Case Control study	W	W	W	NA	S	NA	Weak
45.	Bonyadi et al. [63]	2014	International Journal of Dermatology	Case Control study	W	W	W	NA	S	NA	Weak
46.	Shahneh et al. [64]	2012	Pakistan Journal of Biological Sciences	Cohort study	W	W	M	NA	M	M	Weak
47.	Shi et al. [65]	2014	PLoS ONE	Case control study	W	W	S	W	S	M	Weak
48.	Li et al. [66]	2014	PLoS ONE	Case control study	W	W	S	W	S	M	Weak
49.	Montes-Cano et al. [67]	2013	Arthritis Research & Therapy	Case control study	W	W	S	W	S	M	Weak
50.	Savey et al. [68]	2014	Orphanet Journal of Rare Diseases	Cross Sectional study	W	W	NA	NA	S	NA	Weak
51.	Jiang et al. [69]	2016	Scientific Reports	Case control study	M	W	S	NA	S	M	Moderate
52.	Kawasaki et al. [70]	2010	International Urology and Nephrology	Cross sectional study	W	W	NA	NA	S	NA	Weak

S=STRONG, M=MODERATE, W=WEAK, and NA=NOT APPLICABLE

GLOBAL RATING: STRONG (no WEAK ratings), MODERATE (one WEAK rating), WEAK (two or more WEAK ratings)

Table 2. Summary of extracted data of the selected articles

Sl No.	**Author**	**Year**	**Study design**	**Sample Size**	**Country**	**Types of vasculitis**
53.	Nair et al. [20]	2018	Case Control study	104	India	Takayasu's arteritis
54.	Takamura et al. [21]	2012	Case Control study	96	Japan	Takayasu's arteritis
55.	Terao et al. [22]	2013	Cohort study	379	Japan	Takayasu's arteritis
56.	Gudbrandsson et al. [23]	2016	Cohort study	2800000	Norway	Takayasu's arteritis
57.	Bilge et al. [24]	2012	Cross Sectional study	22	Turkey	Takayasu's arteritis
58.	Gruener et al. [25]	2019	Cohort study	586	USA	Giant cell arteritis
59.	Brekke et al. [26]	2017	Cohort study	792	Norway	Giant cell arteritis
60.	Tan et al. [27]	2019	Cross Sectional study	92	UK	Giant cell arteritis
61.	Pereira et al. [28]	2015	Cross Sectional study	38	USA	Giant cell arteritis
62.	Wang et al. [29]	2019	Case control study	1155	China	Kawasaki disease
63.	Kuo et al. [30]	2011	Case control study	381	Taiwan	Kawasaki Disease
64.	Yeter et al. [31]	2016	Case control study	8748	USA	Kawasaki Disease
65.	Kim et al. [32]	2017	Case series	6	USA	Kawasaki disease
66.	Amano et al. [33]	2019	Clinical trial	82	Japan	Kawasaki Disease
67.	Nagelkerke et al. [34]	2019	Cohort study	4000	UK	Kawasaki disease
68.	Boudiaf et al. [35]	2016	Cross Sectional study	133	Algeria	Kawasaki Disease
69.	Fernandez-Cooke et al. [36]	2019	Cross Sectional study	625	Spain	Kawasaki Disease
70.	Holman et al. [37]	2011	Cross Sectional study	528	USA	Kawasaki disease
71.	Ghimire et al. [38]	2019	Cross Sectional study	10486	USA	Kawasaki disease
72.	Manlhiot et al. [39]	2017	Cross Sectional study	1373	Canada	Kawasaki disease
73.	Nakamura et al. [40]	2011	Cross Sectional study	23337	Japan	Kawasaki disease
74.	Makino et al. [41]	2015	Cross Sectional study	26691	Japan	Kawasaki disease
75.	Makino et al. [42]	2018	Cross Sectional study	31675	Japan	Kawasaki disease
76.	Park et al. [43]	2011	Cross Sectional study	9039	Korea	Kawasaki disease
77.	Karadag et al. [44]	2018	Cohort study	93	UK and Turkey	Polyarteritis Nodosa
78.	Jelusic et al. [45]	2012	Cross sectional study	12	Croatia	Polyarteritis Nodosa
79.	Pearce et al. [18]	2017	Cross sectional study	1217	UK	ANCA Associated Vasculitis

80.	Berti et al. [46]	2017	Cross Sectional study	58	USA	ANCA associated vasculitis
81.	Wu et al. [47]	2015	Case Control study	176	China	ANCA Associated Vasculitis
82.	Fujimoto et al. [48]	2011	Case control study	136	Japan, UK	ANCA Associated Vasculitis
83.	Cao et al. [49]	2011	Case control study	137	USA	ANCA associated vasculitis
84.	Enkhmaa et al. [50]	2015	Case control study	267	USA	ANCA Associated Vasculitis
85.	Ahn et al. [51]	2012	Cohort study	55	Korea	ANCA Associated Vasculitis
86.	Li et al. [52]	2018	Cross Sectional study	43,677,829	China	ANCA Associated Vasculitis
87.	Faurschou et al. [53]	2013	Cross Sectional study	7	Denmark	ANCA Associated Vasculitis
88.	Terrier et al. [54]	2017	Cross Sectional study	760	France	ANCA Associated Vasculitis
89.	Nesher et al. [55]	2016	Cross Sectional study	619	Israel	ANCA Associated Vasculitis
90.	Wada et al. [56]	2012	Cross Sectional study	62	Japan	ANCA Associated Vasculitis
91.	Pearce et al. [57]	2016	Cross sectional study	107	UK	ANCA Associated Vasculitis
92.	Ortiz-Fernadez et al. [58]	2016	Cohort study	404	Spain	Behcet's disease
93.	Saleh et al. [59]	2012	Cross Sectional study	6134	USA	Behcet's disease
94.	Li et al. [60]	2012	Case Control study	722	China	Behcet's disease
95.	Hou et al. [61]	2013	Case Control study	738	China	Behcet's disease
96.	Wu et al. [62]	2014	Case Control study	1086	China	Behcet's disease
97.	Bonyadi et al. [63]	2014	Case Control study	69	Iran	Behcet's disease
98.	Shahneh et al. [64]	2012	Cohort study	290	Iran	Behcet's disease
99.	Shi et al. [65]	2014	Case control study	1617	China	Behcet's Disease
100.	Li et al. [66]	2014	Case control study	1626	China	Behcet's disease
101.	Montes-Cano et al. [67]	2013	Case control study	304	Spain	Behcet's disease
102.	Savey et al. [68]	2014	Cross Sectional study	769	France	Behcet's disease
103.	Jiang et al. [69]	2016	Case control study	384	China	Behcet's disease
104.	Kawasaki et al. [70]	2010	Cross sectional study	120	Japan	Henoch-Schonlein purpura
	Total Participants			**46616166**		

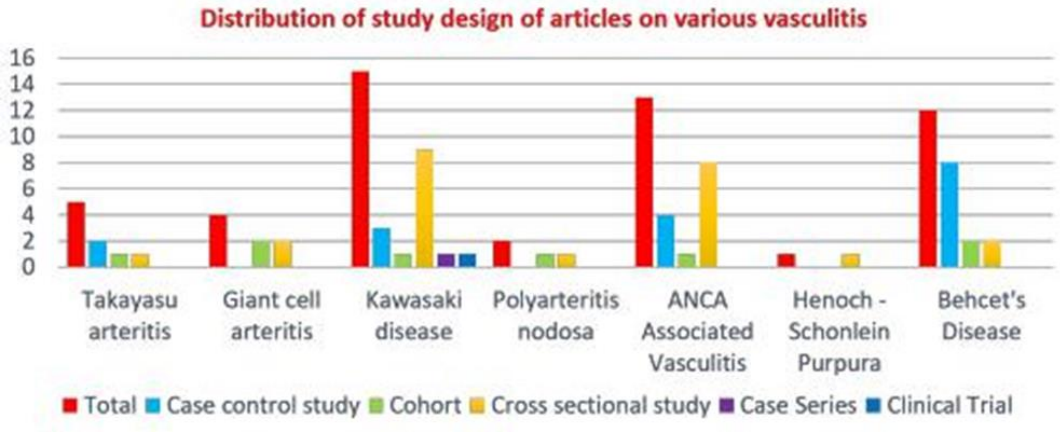

Figure 2. Bar chart illustrating distribution of study designs of articles various vasculitis

Table 3. Association between ethnicity and different types of primary systemic vasculitis

Vessels Size		Vasculitis type	Global Scenario	Ethnic Association			
				Most Common		Least common	
				Ethnic origin	Incidence or Prevalence	Ethnic origin	Incidence or Prevalence
Large sized vessels		Takayasu arteritis [Gudbrandsson et al., 2016; Russo and Katsicas, 2018]	Prevalence: 1-2 per million	Asians	Prevalence: 108 per million	Jews	Prevalence: <1.0 per million
		Giant Cell Arteritis [Richards, March and Gabriel, 2010; Kobayashi and Fujimoto, 2013]	Prevalence: 0.4 to 300 per million	Scandinavians	Prevalence 200 to 320 per million population aged over 50 years	Japanese	Prevalence: 1.47 per 10 million population aged over 50 years
Medium sized vessels		Polyarteritis nodosa [Ungprasert, Koster, Thongprayoon and Warrington, 2016)	Annual incidence: 2 to 30 cases per million	Native Alaskan	Prevalence: 77 per million	South Sweden	Annual incidence 1.6 cases per million
		Kawasaki Disease [Makino et al., 2018; Dietz et al., 2017; [Marchesi et al., 2018]	Annual incidence 34 to 2186 cases per million	Japanese children	Annual incidence: >3000 per million	Native Australian and European children	Annual incidence: 28 to 50 cases per million
Small sized Vessels	ANCA Associated (AAV)	AAV [Pearce et al., 2017; [Berti et a., 2017]	Overall annual incidence: 12-20 per million	Caucasians in Minnesota	Prevalence >400 per million	Hispanic	Annual incidence: 5-15 cases per million
		Granulomatosis with polyangiitis [Greco et al., 2015; Berti et a., 2017; Pierini et al, 2019; Terrier et al., 2017]	Overall prevalence in Europe 50 per million	Caucasians in Minnesota	Prevalence 218 per million	Black sub-Saharans	Annual incidence: 2 to 8 cases per million
		Microscopic polyangiitis [Berti et a., 2017]	Overall Prevalence: 10 to 30 per million	Caucasians in Minnesota	Prevalence 184 per million	Black sub-Saharan	Prevalence: 5 to 10 per million
		Eosinophilic Granulomatosis with polyangiitis (EGPA) [Gokhale et al., 2019; [Berti et a., 2017; Fujimoto et al., 2011]	Overall prevalence in USA 3.2 to 5.9 per million	Caucasians in Minnesota	Prevalence 18 per million and Annual incidence: 4 per million	Unknown	
	Immunocomplex, IgA vasculitis	Henoch-Schonlein Purpura	Annual incidence- Children: 100- 220 cases per million (Piram, Maldini and Mahr, 2012]	Caucasian and Asian children	Annual incidence: 200-250 cases per million	African children	Annual incidence: 2-7 cases per million
Variable sized vessels		Behcet's syndrome [Davatchi et al., 2016]	Annual incidence 5-4200 cases per million	'Silk road' Especially Turkey	Annual Incidence: 200 to 4200 cases per million	United kingdom	Annual incidence: 6.4 cases per million
		Cogan's syndrome	No data found				

Discussion

The incidence and prevalence of vasculitis vary greatly by ethnic group and geographic location. Genetic and environmental factors such as infections, exposure to toxins, and smoking influence the incidence and prevalence of primary systemic vasculitis.

Takayasu's arteritis (TA): TA is a chronic systemic vasculitis affecting large arteries such as the aorta, its major branches, and occasionally coronary and pulmonary arteries [71]. The name "Takayasu disease" was first suggested by Minoru Nakajima in 1921 in the name of great ophthalmologist Professor MikitoTakayasu [72]. In succession, several other names have been proposed, such as (i) avascular disease, (ii) aortic arch syndrome, and (iii) obstructive-producing arteritis [73]. The annual global incidence of TA is estimated at two per million population [8,21]. Russo and co-workers found a higher prevalence of TA in Central and South America, Africans, Indians, and Russians, whereas a rare incidence in Jews [8]. According to Takamura et al. (2012), a higher prevalence of TA has been observed in Southeast Asians, the Indian subcontinent, Brazilians, Chileans, and Argentines compared with other regions [21]. In the southeast Norwegian cohort of 2.8 million population, the point prevalence of Takayasu's arteritis was estimated at 22.0 per million in Northern Europeans, compared with 78.1 per million in Asian whites and 108.3 per million in Africans [23]. An association between TAK and HLA-B52 has been observed in several HLA-B52 ethnic populations [20-24]. Therefore, it is speculated that the higher rate of TAK in Asians may be reflected by the increased frequency of HLA-B52 in this ethnic group. Takamura and colleagues found a positive HLA B67 and HLA B52 association of approximately 5-fold and 4-fold in Japanese patients with Takayasu's arteritis compared with controls [21]. The fundamental role of IL12B in the pathogenesis of HLA-B52associated TA has been described [22]. The increased rate of TA in Asians may reflect a higher frequency of HLA-B52 in this population [24]. For example, the frequency of HLA-B52 in the Japanese population is estimated to be 10%, and the prevalence of TAK is estimated to be 40 per million of the population in Japan. While the frequency of HLA-B52 is estimated to be 2% in the European population, and the incidence of TAK is much lower than in Japan [20-24].

Giant cell arteritis: The most common primary systemic vasculitis in adults over 50 is giant cell arteritis (GCA), which affects medium to large arteries and is affected more often in women than in men [74, 75]. In Europe and North America, giant cell arteritis (GCA) is the most common type of vasculitis [76]. GCA is considered a disease of the elderly, mainly between the ages of 61 and 80, with a male-to-female ratio of about 1:2 [76]. Geographic variations, cyclical patterns, and seasonal fluctuations have been observed in the incidence of GCA [77]. A correlation between the peak incidence of GCA and the viral infection outbreak was identified [77]. Geographic variations in the incidence of GCA are due to a combination of genetic and environmental variables [77]. GCA is more common in Caucasians, especially Scandinavians, with an annual incidence of 20 to 32.0 per 100,000 population, whereas GCA is rare in African Americans over 50 years of age [78]. According to Tan and colleagues, among 92 patients with GCA, Caucasian Britons, descendants of the Indian subcontinent, black British, and other citizens were 66.30, 16.30, 9.78, and 2.17%, respectively [27]. Gruener and colleagues studied the ethnic association of GCA in their 10-year cohort, where the temporal biopsy was performed in 586 individuals with GCA in the United States [25]. In this cohort, 65.2, 28.5. 1.4, 1.2, and 3.8% of participants were White, Black, Hispanic, Asian, and of unknown ethnic origin, respectively,while among biopsy-proven GCA, 81.5, 15.2, and 2.3% were White, Black, and unknown ethnic origin, respectively [25]. The biopsy-proven annual incidence of GCA was estimated at 36 and 31 cases per million, respectively, in whites and blacks in this cohort [25]. In addition, Brekke et al. (2017) found a mean crude incidence of 16.7 and a biopsy-proven incidence of 11.2 per 100,000 Scandinavian patients aged more than 50 years in their retrospective cohort study of 40 years' hospital records [26]. In the UK, patients under 65 have the highest number of deaths in the first year after diagnosis [79]. Genetic susceptibility may be

related to racial/ethnic differences in the incidence and prevalence of GCA. The HLA-DR4 polymorphism is less common in the Japanese population [79]. HLA-DRB04 genetic association was observed in GCA. The association of GCA is two to four times more frequent in individuals with positive HLA-DR4 polymorphisms [80].

Polyarteritisnodosa (PAN): Kussmaul and Maier were the first to describe PAN classically in 1866 [44]. PAN is a moderate inflammatory vasculitisof small blood vessels characterized by necrosis and nodules of aneurysms along the vessel wall. Although PAN is relatively rare in children, it is the third most common vasculitis after HenochSchonleinpurpura and Kawasaki disease in children. The highest prevalenceis in males, with a peak occurring in ages 7-11 [81]. PAN was observed to be the least common vasculitis among primary systemic vasculitis in a large multinational vasculitis observational study (DCVAS - Diagnostic criteria and classification of vasculitis) [82, 83]. The annual worldwide incidence of PAN is 2 to 30 per million population [84]. The annual incidence is estimated to be between 2 and 9 cases per million in Europe and the United States [81]. The highest incidence of PAN has been estimated at 77 per million Alaska natives [82]. The annual incidence is estimated to be 1.6 parts per million in the south [84]. In Jerusalem, the average annual incidence of PAN has been recorded as 3.6 per million adults [55]. The prevalence of PAN was estimated at 30.7 per million inhabitants in Paris in 2000. The overall incidence in European descendants was twice that of non-European ancestry in Paris [85]. The overall annual incidence has decreased in Poland. The annual incidence rates were estimated at 3.3 and 1.9 cases per million population in 2008 and 2013 in Poland, respectively [86].

The downward trend may result from a decrease in the prevalence of hepatitis B during this period in Poland. However, PAN may recur in one-third of cases, and case mortality is estimated at 4% in the United Kingdom [87]. The frequency of PAN was only 3.8% in PSV in Croatian children [45]. The link between geographic location, ethnicity, and NAP development is unclear. Karadag and colleagues (2018) found a younger age at the onset of PAN in the Turkish population than in the British in their group [44]. The risk of recurrence is higher in children with severe gastrointestinal lesions, and the duration of remission is prolonged in such cases [86]. The Mediterranean fever gene (MEFV) mutation is an important underlying factor found in Turkey to create an inflammatory environment. It demonstrates an uncanny response to streptococcal infections, ultimately leading to PAN [88].

Kawasaki Disease: Kawasaki disease (KD) was named after a great Japanese physician, Dr. Tomisaku Kawasaki, who described the disorder well in 1961 [89]. KD was first described in Japan and is now labeled worldwide [90]. The cause of Kawasaki disease (KD) is still unknown. Among infants in the developing world, KD is the leading cause of acquired heart disease, mainly affecting small and medium-sized blood vessels [91]. KD is more common in Japanese children [42]. The annual incidence of Kawasaki disease (KD) in Japanese children aged 0–4 years was estimated at 3030 and 3080 per million in 2013 and 2014, respectively [42]. A higher frequency of KD has been found in children of Asian descent in the United States [38]. Holman et al. (2010) described a higher susceptibility of children of Japanese ancestry in Hawaii, where the highest prevalence of KD (2105 per million) was observed in children of Japanese ancestry under 5 years old,while the lowest incidence (137 per million) was observed in white children of the same age [37]. The overall incidence in US children under 5 years of age has been estimated at 250 per million from hospital data. According to Manlhiot et al., 2017, the annual incidence of KD in Canada is 19.6 per 100,000 children under 5 years of age [39]. East Asian children are 10 to 20 times more likely to develop KD than children living in the West [31]. In Korea, the annual incidence of KD is estimated at 113.1 per 100,000 children, increasing daily [43]. Negelkerke and colleagues found strong, weak, and absent linkage disequilibrium (LD) among Europeans, Africans, and Asian descendants, respectively, in their cohort, including 1028 individuals with KD in the UK [34]. There is a linear association in KD risk across ethnicities, especially among Asian, African, Caucasian, and Hispanic children, and exposure to mercury and cadmium from seafood in particular [31]. The recurrence rate of KD in Japan is estimated to be 3% [9]. The recurrence of

KD was greater than five episodes per thousand patient-years of follow-up in Japan, and the most frequent relapses were observed during the first two years of disease onset [9]. The relative risk of KD is a 10-fold increase in individuals with a positive family history compared with the general Japanese population [91]. The recurrence rate of KS in Canada has been estimated at 2.9 episodes per thousand patient-years of follow-up, with recurrence occurring on average 1.5 years after the first episode with similar clinical features. [92]. Evidence for genetic factors for vulnerability to KD includes the following such as (i) observed higher incidence in Japanese children and children of living Japanese ancestry Outside of Japan, (ii) the prevalence of KD is higher in children with a history of KD in their parents, and (iii) the prevalence of KD is higher in siblings and twins of the same parent [9]. Single nucleotide polymorphisms in six genes or genomic regions such as (i) FcyR2a, (ii) caspase 3 (PASP3), (iii) human leukocyte antigen (HLA) class II, (iv) Bcell lymphoid kinase (BLK), (v) inositol 1,4,5 triphosphate kinase-C (ITPKC) and (vi) CD40 have been described in genome-wide association and family studies [9]. In children of European ancestry, a higher risk of coronary aneurysm was associated with variants of the TGFβ2, TGFβR2, and SMAD3 genes [9]. An association between the HLA determinant and susceptibility to KD was observed in Japanese and Taiwanese children. Nonetheless, no association between HLA determinants and susceptibility to KD was found in European- ancestry children [9]. Higher KD occurrence was found in winter and spring compared with fall, and there is a strong association between first- and second-degree relatives with KD history [36]. Seasonal peaks of KD are observed in winter in the tropical northern hemisphere. On the other hand, low numbers are observed in autumn and summer. However, no seasonal variation of KD was observed in the tropical and extratropical south hemispheres [9].

ANCA-Associated Vasculitis: ANCA-associated vasculitis (AAV) has been well recognized since 1948 [93]. ANCA-associated vasculitis (AAV) is a group of primary systemic vasculitis that affects small blood vessels and causes vascular necrosis in the presence of ANCA in the blood. AAV is classified into three types (i) microscopic polyangiitis (MPA), (II) granulomatosis with polyangiitis (GPA), and (iii) Eosinophilicgranulomatosis with polyangiitis (EGPA) [93]. The annual incidence of ANCAassociated vasculitis was estimated at 23.1 per million in the UK cohort [57]. The annual incidence of AAV was 25.8 per million in whites, whereas 8.4/1000 in blacks and minority ethnic (BME) populations [57]. The annual incidence of AAV among Jews is estimated at 8 per million population [55]. Fujimoto and co-workers found no difference between the UK and Japan, with annual incidence rates estimated at 21.8 and 22.6 per million adults in the UK and Japan, respectively [48]. The overall prevalence of AAV has been reported to be 42.1 per 100,000, with an average annual incidence of 3.3 per 100,000 population in Minnesota, USA. An incidence of AAV was found in 0.25% of Dong, Zhuang, and Li Chinese ethnic populations, with a positive correlation between carbon monoxide exposure and AAV development [52]. Furthermore, the overall incidence of AAV has been estimated at 131–183, 198, 137, and 209 per 10 million people per year in Spain, the United Kingdom, Australia, and Sweden, respectively. GPA is more common than MPA in northern Europe, and MPA is more common than GPA in southern Europe [94,95]. In the United Kingdom, the combined annual incidence of GPA and MPA was estimated at 1.5 cases per million cases in the 1980s [96]. The overall prevalence of AAV ranges from 30 to 218 cases per million population worldwide [96]. The literature suggests that the prevalence of AAV has increased in recent years as both survival and incidence of AAV have increased [96]. For example, AAV prevalence in northern and southern Germany was 74 per million in 1994 and 149 per million in 2006, only doubling in 12 years. Katsuyama and co-workers reported ANCA positive for MPO was higher in Japan (83.7%) than in the UK (30.0%) [17]. On the other hand, PR3-positive AAV was reported to be lower in Japan (7.0%) compared to the UK (58.0%) [17]. The estimated annual incidence of total AAV in children was reported at 0.22 to 0.45 per million and 3.2 per million in French and Sweden [96]. Pearce and colleagues reported that people from Northern Europe, Turkey, and the Indian subcontinent had higher PR3-ANCA-associated

vasculitis [57]. Japanese and Chinese have a 60fold and 7-fold higher risk of developing MPOANCA-associated vasculitisthan northern Europeans [48]. A higher likelihood of MPOAAV is also observed in Caucasian Americans, Turks, and Southern Europeans compared to Northern Europeans. A higher proportion of MPO-positive AAV patients than PR3-positive AAV has also been reported in Asian countries such as China and Taiwan [48]. Sreihet and colleagues reviewed disease severity in Caucasian and Hispanic patients with AAV 2014 [12]. Hispanic patients have been reported to exhibit more severe disease activity and greater organ involvement than Caucasians residing in the same geographic area [12, 97]. Both type I and type II HLA genes are predisposed to developing AAV. HLA-B*08, HLA-A*01, and HLA-DRB1*03 have been reported to be genetically susceptible to AAV, especially GPA [98]. The relationship between HLA class II genes and clinically apparent disorders or ANCA archetypes fluctuates between ethnicity and geographic location. HLADRB1*13 was protective against PR3-ANCApositive Dutch GPA and German GPA patients [98]. While HLA-DRB1*04 was found to be associated with Dutch GPA patients and German GPA-positive ANCA PR3 patients with an increased risk of developing end-stage renal disease (ESRD) [98]. In addition, among British and German patients with GPA, HLA-DBP1*04 was reported more frequently. In Caucasians, the HLA-DBP1 polymorphism, rs3117242(G), has been reported to confer genetic susceptibility to the development of GPA, especially PR3-ANCApositive GPA [98]. Among African Americans and Chinese, the genetic factors HLA-DRB1*15 and HLA-DBR1*1202 have been reported to be susceptible to PR3-ANCA-positive vasculitis, respectively. Japanese patients with MPA-positive AAV and MPO have been reported to be associated with HLA-DRB1*0901 and its haplotype [98]. Prognosis and disease severity are also associated with HLA polymorphisms. Poor prognosis and poor response to treatment have been reported in HLA-DRB1*0405-positive patients, while higher mortality has been reported in Chinese AAV patients with HLA-DRB1*0403 [98]. Interestingly, the HLADPB1/RXRB*0301/01 haplotype was associated with a lower risk of GPA, while the HLADPB1/RXRB*0401/03 haplotype was associated with a higher risk of GPA. The RXRB rs6531 polymorphism was commonly found in patients with a positive mean GPA score for ANCA. Ring finger protein-1 (RING) polymorphisms such as rs213210, rs213209, and rs213208 are associated with GPA patients in Germany [98]. Severe symptoms of GPA were observed in white Europeans compared with sub-Saharan Africans and Afro-Caribbeans (hazard ratio 1.96) [54]. The prevalence of GPA increased from 28.8 to 64.8 per million UK population in 2020 [96]. In southern Sweden, the prevalence of GPA was reported to be 160 per million, much higher than in the UK. Similarly, in the United States, a survey of insurance claims databases found the prevalence of GPA to be 30.5 per million adults aged 18 to 65 [99]. However, the highest prevalence of her GPA in the United States (218 per million) was reported in his 20-year study conducted at the Mayo Clinic in the United States [46]. Methodological differences between studies may be a possible reason for this large variation in the prevalence of GPA in the United States. Alike USA, the prevalence is also rising in Norway [85]. In 1988 his GPA was 30.4 per million, while in 1998, he was 95.1 per million in Norway [85]. This is a more than three-fold increase in prevalence during his ten years in Norway [85]. In New Zealand and Australia, the prevalence of GPA is estimated at 112 and 95 per million, respectively [99]. Similar to total AAV, a trend of increasing incidence of GPA has been observed worldwide [99].

Eosinophilic granulomatosis with polyangiitis (EGPA), also known as Churg-Strauss disease, was developed by two physicians, Dr. Churg and Dr. Strauss, who described EGPA in detail in 1951 as eosinophil-rich necrotic granulomatous inflammation of small to medium-sized vessels. EPGA commonly affects the airways and is associated with the presence of asthma. The coexistence of glomerulonephritis and ANCA is common in EGPA. EGPA has the highest prevalence in Australia, estimated at 22.3 per million population [96]. The prevalence of EGPA was given as 10.7, 13, 14, and 18 cases per million population in Paris, Norway, Sweden, and the United States, respectively [46, 96]. The annual incidence of EGPA in children aged 0 to 17 years

is estimated at 0.4 cases per million in Sweden [96].

Immune complex microvasculitis (ISVV): is the deposition of immunoglobulins and/or complement components in the walls of small vessels in ISVV [3]. Immune complex vasculitis includes (i) Henoch-Schönlein purpura (HSP/IgA vasculitis), (ii) anti-glomerular basement membrane (anti-GBM) disease, (iii) cryoglobulinemic vasculitis, and (iv) hypocomplementaemic urticaria vasculitis (HUV/anti-C1q vasculitis) [3]. The most common form of vasculitis in children is Henoch Schönlein purpura (HSP), also known as immunoglobulin A vasculitis (IgAV) [100]. IgAV usually presents as an acute non thrombocytopenic vasculitis affecting small blood vessels before 10 years, with peak onset between 4 and 6 years of age and a slight predominance in males. However, IgAV is very rare in infants. The global incidence of IgAV has been estimated at 30–270 cases per million children per year [100]. In most ethnic groups in Europe, the annual incidence is estimated at 220 per million people under 14 [16]. Asian ethnic groups, especially Korean and Japanese children, are strongly affected by her IgAV compared to African ancestry [100]. The average annual incidence of HSP nephritis in Japan is 34–36 per million pediatric population [70]. However, HSP is less than one-third in black children compared to Asian or white children [16]. Moreover, hospitalization rates have been reported among ethnic groups, such as 1.9 Hispanics, 1.6 Whites, and 0.9 Blacks per 100,000 children in the United States [101]. Because viral infections can cause IgAV, seasonal variations in outbreaks have been shown [100]. In South Korea, the seasonal variation of IgAV in 16,000 children was studied, with fewer cases in summer than in winter and autumn [101]. HSP susceptibility is strongly associated with HLA-DRB1*0103 [102]. However, non-HLA genetic markers can also influence HSP susceptibility and disease severity [102].

Anti-Glomerular Basement Membrane (AntiGBM) Diseases: Anti-GBM disease or 'Goodpasture's disease' has been described as a rare type of PSV that affects renal and/or pulmonary capillaries. The overall annual incidence of the anti-GBM disease is estimated to be less than 1 per million European population. In Ireland, the estimated incidence of the disease was 1.6 per million population per year. Anti-GBM disease was well recognized among Asians and Caucasians but was considered rare in Africa [103]. The literature describes seasonal variations and outbreaks, particularly the temporal and spatial accumulation of disease. Anti-GBM disease is caused by environmental factors such as influenza infection and smoking [103]. Genetic factors predisposing to anti-GBM disease may be triggered by environmental factors. The HLADR2 haplotype is estimated to be inherited 80% of the time. HLA-DBR1 alleles, specifically HLADRB1*1501 and HLA-DRB1*0401, have been reported to be positively associated with this disease, whereas HLA-DRB1*07 is associated with this diseaseand showed a protective effect. HLA-DRB1*1501 associations have been reported in Asian populations [103]. In addition, susceptibility to diseases of non-HLA gene polymorphisms has also been reported. Non-HLA genes encoding Fcγ receptors and COL4A3 polymorphisms have been described in disease manifestation [103].

Cryoglobulinemia: Cryoglobulinemia is characterized by the constant presence of abnormal immunoglobulins (Ig) in the serum, which accelerates at low temperatures and decays again with warming [1]. Cryoglobulinemia consists of (i) type I cryoglobulinemia- consisting of monoclonal immunoglobulin (Ig), (ii) type II cryoglobulinemia- consisting of monoclonal Ig binding to polyclonal heavy chains, and (iii) III Type cryoglobulinemia - is divided into three types consisting of polyclonal immunoglobulins. Cryoglobulinemia is a rare disease in the general population of Europe and the United States. The overall prevalence of cryoglobulinemia is estimated at < 5 per 10,000 populations in the United States and Europe [105]. However, a higher prevalence has been reported in the Mediterranean region. Mixed cryoglobulinemia, especially type II, can be caused by hepatitis-C virus (HCV) infection. Despite geographical differences, HCV infection is widespread in Central Asia, East Asia, and Northeast Africa and is estimated to infect more than 184 million people worldwide. The estimated prevalence of HCV in

the above regions ranged from 15 to 35 per 1,000 population, with a prevalence of less than 1/2 in Western Europe, the United Kingdom, Asia Pacific, North America, and Latin America [105]. It is estimated at 10-20 in the Mediterranean region and 25-30 per 1,000 inhabitants in Eastern Europe [105]. Several studies have reported that cryoglobulinemia occurs in 10-60% of HCVinfected cases [104 -106]. A genome-wide association study (GWAS) shows a significant association between cryoglobulin-associated vasculitis (CryVas) and a single nucleotide polymorphism (SNP, rs9461776) near NOTCH4 and HLA-DRB1 and HLA-DQA1 on chromosome 6 reported [106].

Hypocomplement Urticaria-like vasculitis (HUV/anti-C1q vasculitis): HUV is a form of immune complex small vessel vasculitis, characterized by urticaria and hypocomplement-emia for at least 6 months, accompanied by (i) arthralgia, (ii) uveitis, (iii) glomerulonephritis, and/or (iv) characterized by comorbid systemic symptoms such as recurrent abdominal pain [107]. In addition, chronic obstructive pulmonary disease (COPD) is commonly found in his HUV and is the leading cause of HUV mortality and morbidity. Global incidence and prevalence have not been reported to date. However, the annual incidence in Sweden is estimated at 7 per 10 million people. His estimated point prevalence of HUV in Sweden in 2015 was 9.5 per million. In addition, 57 cases of HUV were recorded in the French national database [107].

Behcet's disease: Behcet's disease (BD) or Adamantiades-Behcet's disease is an idiopathic, chronic, recurrent vasculitis that can involve the mucocutaneous, joints, blood vessels, eyes, gastrointestinal tract, and central nervous system (CNS) affecting by variable vessel vasculitis, including arteries, veins, arterioles, venules, and capillaries [3]. Mortality and morbidity are significantly higher in BD. It primarily affects young adults due to its geographic distribution known as the "Silk Road", which corresponds to ancient routes between the Middle East, Mediterranean, andthe Far East. BD has the highest prevalence in Turkey,with 3700 per million population. The prevalence of BD in Japan is estimated at 160 per million people, and more than 20,000 patients sought medical help in Japan in 2014 [108]. Low incidence and prevalence have been reported in Europe and North America. Brazil, Africa, and Australia have also reported a few cases. Interestingly, the incidence of BD is lower among Armenians living in Istanbul, Turkey [1]. Phenotypic representation among different ethnic groups can vary in BD. Patients with BD in sub-Saharan Africa have been reported to have greater central nervous system involvement, higher mortality, and lower frequencies of the HLA-B51 allele compared with those in North Africa and Europe, respectively. [68]. A genetic association has been reported in BD [108, 109]. Significant associations of HLAB5, HLA-B35, HLA-51, HLA-B52, and HLACW4 in BD patients have been reported in the Iranian Azerbaijani population [64, 109]. However, while there is no statistically significant association between the PTPN22 gene and BD in the Spanish population [56], there is a significant association between the PTPN2 polymorphism and BD in the Han population [62]. Nonetheless, no significant association of his MDR1C3435T polymorphism was found between a BD patient and his Turkish healthcare manager in Azerbaijan, Iran [63]. The rs9494885 TC genotype is common twice in BD patients compared with controls [60]. However, HLA-B51 is found in 15% of the Japanese population, whereas one-third of Behcet's disease is not associated with HLA-B51 [108].

Cogan Syndrome: Cogan syndrome (CS) is a rare autoimmune vasculitis [110]. The main features of CS are eye inflammation and sensorineural hearing loss. Young adults are more commonly affected, but people of all ages can suffer from the disease. Anti-Hsp70 antibodies are associated with CS. It can occur in individuals of any race or ethnic group but is more common in Caucasians [111]. CS is one of the rarest PSVs, has no gender predominance, and affects only young adults with a peak age of 29 [112]. Approximately 250 cases have been reported so far, most of them Caucasian [112]. In expert opinion, the actual number of cases of CS is dwarfed by (i) idiopathic recurrent keratitis, (ii) idiopathic deafness, and (iii) underdiagnoses or lack of diagnosis as an autoimmune inner ear

disease. It has been explained that there may be more cases than published [112].

Single Organ Vasculitis: Instead of "focal vasculitis", the term single organ vasculitis (SOV) is considered more appropriate. SOV is self-evident vasculitis confined to a single organ, such as the skin, central nervous system, or reproductive tract. SOV is classified into three subclasses: (i) focal single-organ vasculitis, (ii) multiple single-organ vasculitis, and (iii) diffuse single-organ vasculitis [113]. However, SOV has an inherent risk of conversion to systemic vasculitis, and continuous follow-up for at least 6 months should be considered and implemented. Primary central nervous system vasculitis (PCNS) is SOV's most commonly reported form. Approximately 500 cases of PCNS vasculitis have been reported [113]. PCNS vasculitis in the United States is 2.4 cases per million yearly population. Non systemic vasculitic neuropathy (NSVN), limited renal vasculitis, pulmonary Takayasu disease, retinal vasculitis, thoracic vasculitis, vasculitis of genitourinary structures, gallbladder vasculitis, and aortic vasculitis are described in case reports [113]. However, no data were found on ethnic associations in occurrence in geographical locations.

Limitations: Articles related to PSV published in the last 10 years are included in this review. In addition, a systematic search of English literature was conducted in the PubMed and Google Scientist databases. Therefore, some published articles may not be included in this review. Due to differences in study design, we could not perform a meta-analysis. Therefore, pooled prevalence and incidence of primary systemic vasculitis were not estimated for any ethnic group in this review.

Conclusion

Although evidence for an association between ethnicity and different types of vasculitis is limited, the prevalence and incidence of PSV vary widely among different ethnic and/or racial groups worldwide.Genetic susceptibility and environmental factors also contribute to the incidence and prevalence of primary systemic vasculitis.

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References

1. Bijlsma J. EULAR Textbook On Rheumatic Diseases. 1st ed. London: BMJ Publishing Group. 2013: 616-636.
2. Types of Vasculitis: Johns Hopkins Vasculitis Center. Johns Hopkins Vasculitis Center. 2022. https://www.hopkinsvasculitis.org/types-vasculitis/ (Accessed 29 October 2022).
3. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis & Rheumatism. 2012; 65(1):1–11.
4. van der Geest KSM, Sandovici M, van Sleen Y, Sanders J-S, Bos NA, Abdulahad WH, Stegeman CA, Heeringa P, et al. Review: What Is the Current Evidence for Disease Subsets in Giant Cell Arteritis? Arthritis & Rheumatology. 2018; 70(9):1366–1376.
5. Imai N, Kuroda R, Konishi T, Serizawa M, Kobari M. Giant Cell Arteritis: Clinical Features of Patients Visiting a Headache Clinic in Japan. Internal Medicine. 2011; 50(16):1679–1682.
6. Crow RW, Katz BJ, Warner JEA, Alder SC, Zhang K, Schulman S, Digre KB. Giant Cell Arteritis and Mortality. The Journals of Gerontology Series A: Biological Sciences and Medical Sciences. 2009; 64A(3):365–369.
7. Comarmond C, Biard L, Lambert M, Mekinian A, Ferfar Y, Kahn J-E, et al. Long-Term Outcomes and Prognostic Factors of Complications in Takayasu Arteritis. Circulation. 2017; 136(12):1114–1122.
8. Russo RAG, Katsicas MM. Takayasu Arteritis. Frontiers in Pediatrics. 2018; 6.
9. McCrindle BW, Rowley AH, Newburger JW, Burns JC, Bolger AF, Gewitz M, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. Circulation. 2017; 135(17).
10. Dietz SM, van Stijn D, Burgner D, Levin M, Kuipers IM, Hutten BA, Kuijpers TW. Dissecting Kawasaki disease: a state-of-the-art review. European Journal of Pediatrics. 2017; 176(8):995–1009.
11. Hoang MP, Park J. Vasculitis. Hospital-Based Dermatopathology. 2020; 245–296.
12. Sreih AG, Annapureddy N, Springer J, Casey G, Byram K, Cruz A, et al. Development and validation of case-finding algorithms for the identification of patients with anti-neutrophil cytoplasmic antibody-associated vasculitis in large healthcare administrative databases. Pharmacoepidemiology and Drug Safety. 2016; 25(12):1368–1374.
13. Demirkaya E, Arici ZS, Romano M, Berard RA, Aksentijevich I. Current State of Precision Medicine in Primary Systemic Vasculitides. Frontiers in Immunology. 2019; 10.

14. Bonatti F, Reina M, Neri TM, Martorana D. Genetic Susceptibility to ANCA-Associated Vasculitis: State of the Art. ***Frontiers in Immunology*** 2014; 5:1-14.
15. Brogan P, Eleftheriou D. Vasculitis update: pathogenesis and biomarkers. ***Pediatric Nephrology*** 2017; 33(2):187–198.
16. Piram M, Maldini C, Mahr A. Effect of race/ethnicity on risk, presentation, and course of connective tissue diseases and primary systemic vasculitides. ***Current Opinion in Rheumatology***. 2012; 24(2):193–200.
17. Katsuyama T, Sada K-E, Makino H. Current Concept and Epidemiology of Systemic Vasculitides. Allergology International. 2014; 63(4):505–513.
18. Pearce FA, Craven A, Merkel PA, Luqmani RA, Watts RA. Global ethnic and geographic differences in the clinical presentations of anti-neutrophil cytoplasm antibody–associated vasculitis. Rheumatology. 2017; 56(11):1962–1969.
19. Berti A, Dejaco C. Update on the epidemiology, risk factors, and outcomes of systemic vasculitides. Best Practice & Research Clinical Rheumatology. 2018; 32(2):271–294.
20. Nair AM, Goel R, Hindhumati M, Shah K, Chandana P, Jayaseelan V, et al. C-reactive protein gene polymorphisms (rs1205) in Asian Indian patients with Takayasu arteritis: Associations and phenotype correlations. International Journal of Rheumatic Diseases. 2017; 21(3):732–739.
21. Takamura C, Ohhigashi H, Ebana Y, Isobe M. New Human Leukocyte Antigen Risk Allele in Japanese Patients With Takayasu Arteritis. Circulation Journal. 2012; 76(7):1697–1702.
22. Terao C, Yoshifuji H, Kimura A, Matsumura T, Ohmura K, Takahashi M, et al. Two Susceptibility Loci to Takayasu Arteritis Reveal a Synergistic Role of the IL12B and HLA-B Regions in a Japanese Population. The American Journal of Human Genetics. 2013; 93(2):289–297.
23. Gudbrandsson B, Molberg Ø, Garen T, Palm Ø. Prevalence, Incidence, and Disease Characteristics of Takayasu Arteritis by Ethnic Background: Data From a Large, Population-Based Cohort Resident in Southern Norway. Arthritis Care & Research. 2016; 69(2):278–285.
24. Bilge NSY, Kaşifoğlu T, Cansu DÜ, Korkmaz C. Retrospective evaluation of 22 patients with Takayasu's arteritis. Rheumatology International. 2011; 32(5):1155–1159.
25. Gruener AM, Poostchi A, Carey AR, Eberhart CG, Henderson AD, Chang JR, McCulley TJ. Association of Giant Cell Arteritis with Race. JAMA Ophthalmology. 2019; 137(10):1175.
26. Brekke LK, Diamantopoulos AP, Fevang B-T, Aβmus J, Esperø E, Gjesdal CG. Incidence of giant cell arteritis in Western Norway 1972–2012: a retrospective cohort study. Arthritis Research & Therapy. 2017; 19(1).
27. Tan N, Acheson J, Ali N. Giant cell arteritis in patients of Indian Subcontinental descent in the UK. Eye. 2018; 33(3):459–463.
28. Pereira LS, Yoon MK, Hwang TN, Hong JE, Ray K, Porco T, McCulley TJ. Giant cell arteritis in Asians: a comparative study. British Journal of Ophthalmology. 2010; 95(2):214–216.
29. Wang Y, Lu Z, Fu L, Tan Y, Che D, Huang P, Pi L, Xu Y, Liang Q, Zhang L, Qiu X, Gu X. The miRNA-608 rs4919510 G>C polymorphism confers reduce coronary injury of Kawasaki disease in a Southern Chinese population. Bioscience Reports. 2019; 39(5):1-7.
30. Kuo H-C, Onouchi Y, Hsu Y-W, Chen W-C, Huang J-D, Huang Y-H, Yang Y-L, Chao M-C, Yu H-R, Juan Y-S, Kuo C-M, Yang KD, Huang J-S, Chang W-C. Polymorphisms of transforming growth factor-β signaling pathway and Kawasaki disease in the Taiwanese population. Journal of Human Genetics. 2011; 56(12):840–845.
31. Yeter D, Portman M, Aschner M, Farina M, Chan W-C, Hsieh K-S, Kuo H-C. Ethnic Kawasaki Disease Risk Associated with Blood Mercury and Cadmium in U.S. Children. International Journal of Environmental Research and Public Health. 2016; 13(1):101.
32. Kim J, Shimizu C, Kingsmore SF, Veeraraghavan N, Levy E, Ribeiro dos Santos AM, Yang H, Flatley J, Hoang LT, Hibberd ML, Tremoulet AH, Harismendy O, Ohno-Machado L, Burns JC. Whole genome sequencing of an African American family highlights toll-like receptor 6 variants in Kawasaki disease susceptibility. Prokunina-Olsson L, ed. PLOS ONE. 2017; 12(2):e0170977.
33. Amano Y, Akazawa Y, Yasuda J, Yoshino K, Kojima K, Kobayashi N, Matsuzaki S, Nagasaki M, Kawai Y, Minegishi N, Ishida N, Motoki N, Hachiya A, Nakazawa Y, Yamamoto M, et al. A low-frequency IL4R locus variant in Japanese patients with intravenous immunoglobulin therapy-unresponsive Kawasaki disease. Pediatric Rheumatology. 2019; 17(1).
34. Nagelkerke SQ, Tacke CE, Breunis WB, Tanck MWT, Geissler J, Png E, et al. Extensive Ethnic Variation and Linkage Disequilibrium at the FCGR2/3 Locus: Different Genetic Associations Revealed in Kawasaki Disease. Frontiers in Immunology. 2019; 10.185.
35. Boudiaf H, Achir M. The Clinical Profile of Kawasaki Disease in Algerian Children: A Single Institution Experience. Journal of Tropical Pediatrics. 2015; 62(2):139–143.
36. Fernandez-Cooke E, Barrios Tascón A, Sánchez-Manubens J, Antón J, Grasa Lozano CD, Aracil Santos J, Villalobos Pinto E, Clemente Garulo D, Mercader Rodríguez B, Bustillo Alonso M, NuñezCuadros E, et al. Epidemiological and clinical features of Kawasaki disease in Spain over 5 years and risk factors for aneurysm development. (2011-2016): KAWA-RACE study group. Corsini I, ed. PLOS ONE. 2019; 14(5):e0215665.
37. Holman R, Christensen K, Belay E, Steiner C, Effler P, Miyamur J, Forbes S, Schonberger L, Melish M. Racial/Ethnic Differences in the Incidence of Kawasaki Syndrome among Children in Hawaii. Hawaii Medical Journal. 2010; 69(8): 194-197.
38. Ghimire LV, Chou F-S, Mahotra NB, Sharma SP. An update on the epidemiology, length of stay, and cost of Kawasaki disease hospitalization in the United States. Cardiology in the Young. 2019; 29(6):828–832.
39. Manlhiot C, O'Shea S, Bernknopf B, LaBelle M, Chahal N, Dillenburg RF, Lai LS, Bock D, Lew B, Masood S, Mathew M, McCrindle BW. Epidemiology of Kawasaki Disease in Canada 2004 to 2014: Comparison of Surveillance Using Administrative Data vs Periodic

Medical Record Review. Canadian Journal of Cardiology. 2018; 34(3):303–309.

40. Nakamura Y, Yashiro M, Uehara R, Sadakane A, Tsuboi S, Aoyama Y, Kotani K, Tsogzolbaatar E-O, Yanagawa H. Epidemiologic Features of Kawasaki Disease in Japan: Results of the 2009-2010 Nationwide Survey. Journal of Epidemiology. 2012; 22(3):216–221.
41. Makino N, Nakamura Y, Yashiro M, Ae R, Tsuboi S, Aoyama Y, Kojo T, Uehara R, Kotani K, Yanagawa H. Descriptive Epidemiology of Kawasaki Disease in Japan, 2011-2012: From the Results of the 22nd Nationwide Survey. Journal of Epidemiology. 2015; 25(3):239–245.
42. Makino N, Nakamura Y, Yashiro M, Sano T, Ae R, Kosami K, Kojo T, Aoyama Y, Kotani K, Yanagawa H. Epidemiological observations of Kawasaki disease in Japan, 2013-2014. Pediatrics International. 2018; 60(6):581–587.
43. Park YW, Han JW, Hong YM, Ma JS, Cha SH, Kwon TC, Lee SB, Kim CH, Lee JS, Kim CH. Epidemiological features of Kawasaki disease in Korea, 2006-2008. Pediatrics International. 2011; 53(1):36–39.
44. Karadag O, Erden A, Bilginer Y, Gopaluni S, Sari A, Armagan B, Ertenli I, Ozen S, Jayne D. A retrospective study comparing the phenotype and outcomes of patients with polyarteritis nodosa between UK and Turkish cohorts. Rheumatology International. 2018; 38(10):1833–1840.
45. Jelusic M, Vikic-Topic M, Batinic D, Milosevic D, Malenica B, Malcic I. Polyarteritis nodosa in Croatian children: a retrospective study over the last 20 years. Rheumatology International. 2012; 33(12):3087–3090.
46. Berti A, Cornec D, Crowson CS, Specks U, Matteson EL. The Epidemiology of Antineutrophil Cytoplasmic Autoantibody-Associated Vasculitis in Olmsted County, Minnesota. Arthritis & Rheumatology. 2017; 69(12):2338–2350.
47. Wu Z, Wu Q, Xu J, Chen S, Sun F, Li P, Bai Y, Zheng W, Chen H, Zhang F, Li Y. HLA-DPB1 variant rs3117242 is associated with anti-neutrophil cytoplasmic antibody-associated vasculitides in a Han Chinese population. International Journal of Rheumatic Diseases. 2015; 20(8):1009–1015.
48. Fujimoto S, Watts RA, Kobayashi S, Suzuki K, Jayne DRW, Scott DGI, Hashimoto H, Nunoi H. Comparison of the epidemiology of anti-neutrophil cytoplasmic antibody-associated vasculitis between Japan and the UK. Rheumatology. 2011; 50(10):1916–1920.
49. Cao Y, Schmitz JL, Yang J, Hogan SL, Bunch D, Hu Y, Jennette CE, Berg EA, Arnett FC, Jennette JC, Falk RJ, Preston GA. DRB1*15Allele Is a Risk Factor for PR3-ANCA Disease in African Americans. ***Journal of the American Society of Nephrology*** 2011; 22(6):1161–1167.
50. Enkhmaa B, Anuurad E, Zhang W, Kim K, Berglund L. Diverging trajectory patterns of systemic versus vascular inflammation over age in healthy Caucasians and African-Americans. ***Atherosclerosis*** 2015; 239(2):509–515.
51. Ahn JK, Hwang J-W, Lee J, Jeon CH, Cha H-S, Koh E-M. Clinical features and outcome of microscopic polyangiitis under a new consensus algorithm of ANCA-associated vasculitides in Korea. ***Rheumatology International*** 2011; 32(10):2979–2986.
52. Li J, Cui Z, Long J, Huang W, Wang J, Wang H, Zhang L, Chen M, Zhao M. The frequency of ANCA-associated vasculitis in a national database of hospitalized patients in China. Arthritis Research & Therapy. 2018; 20(1).
53. Faurschou M, Helleberg M, Obel N, Baslund B. Incidence of granulomatosis with polyangiitis (Wegener's) in Greenland and the Faroe Islands: epidemiology of an ANCA-associated vasculitic syndrome in two ethnically distinct populations in the North Atlantic area. Clin Exp Rheumatol. 2013; 31(75):S52-S55.
54. Terrier B, Dechartres A, Deligny C, Godmer P, Charles P, Hayem G, et al. Granulomatosis with polyangiitis according to geographic origin and ethnicity: clinical-biological presentation and outcome in a French population. Rheumatology. 2016:kew423.
55. Nesher G, Ben-Chetrit E, Mazal B, Breuer GS. The Incidence of Primary Systemic Vasculitis in Jerusalem: A 20-year Hospital-based Retrospective Study. The Journal of Rheumatology. 2016; 43(6):1072–1077.
56. Wada T, Hara A, Arimura Y, Sada K-E, Makino H. Risk Factors Associated with Relapse in Japanese Patients with Microscopic Polyangiitis. The Journal of Rheumatology. 2011; 39(3):545–551.
57. Pearce FA, Lanyon PC, Grainge MJ, Shaunak R, Mahr A, Hubbard RB, Watts RA. Incidence of ANCA-associated vasculitis in a UK mixed ethnicity population. Rheumatology. 2016; 55(9):1656–1663.
58. Ortiz-Fernández L, Montes-Cano M, García-Lozano J, Conde-Jaldón M, Ortego-Centeno N, González-Leon R, et al. PTPN22 is not associated with Behçet's disease. A study spanning the complete gene region in the Spanish population and meta-analysis of the functional variant R620W. Clinical and Experimental Rheumatology. 2016; 34(6):1-6.
59. Saleh OA, Birnbaum AD, Tessler HH, Goldstein DA. Behçet Uveitis in the American Midwest. Ocular Immunology and Inflammation. 2012;20(1):12–17.
60. Li H, Liu Q, Hou S, Du L, Zhou Q, Zhou Y, Kijlstra A, Li Z, Yang P. TNFAIP3 gene polymorphisms confer risk for Behcet's disease in a Chinese Han population. Human Genetics. 2012; 132(3):293–300.
61. Hou S, Qi J, Zhang Q, Liao D, Li Q, Hu K, Zhou Y, Kijlstra A, Yang P. Genetic variants in the JAK1 gene confer a higher risk of Behcet's disease with ocular involvement in Han Chinese. Human Genetics. 2013; 132(9):1049–1058.
62. Wu Z, Chen H, Sun F, Xu J, Zheng W, Li P, et al. PTPN2 rs1893217 single-nucleotide polymorphism is associated with risk of Behçet's disease in a Chinese Han population. Clin Exp Rheumatol. 2014; 32(84):S20-S26.
63. Bonyadi M, Gholizadeh M, Soltan-Ali M. MDR1C3435T polymorphism associated with the development of clinical features in Behçet's disease in Iranian Azeri Turkish patients. International Journal of Dermatology. 2014; 53(10):1235–1240.
64. Shahneh FZ, Babaloo Z, Baradaran B, Hamzavi F, Bayazi B, Bandehagh A. Behçet's Syndrome in Iranian Azari People. Pakistan Journal of Biological Sciences. 2012; 15(21):1045–1047.
65. Shi Y, Jia Y, Hou S, Fang J, Zhou Y, Kijlstra A, Yang P. Association of a TNIP1 Polymorphism with Vogt-

Koyanagi-Harada Syndrome but Not with Ocular Behcet's Disease in Han Chinese. Anderson MG, ed. PLoS ONE. 2014;9(5):e95573.

66. Li X, Bai L, Fang J, Hou S, Zhou Q, Yu H, Kijlstra A, Yang P. Genetic Variations of IL-12B, IL-12Rβ1, IL-12Rβ2 in Behcet's Disease and VKH Syndrome. Yao Y-G, ed. PLoS ONE. 2014;9(5):e98373.
67. Montes-Cano M, Conde-Jaldón M, García-Lozano J, Ortiz-Fernández L, Ortego-Centeno N, Castillo-Palma M, et al. HLA and non-HLA genes in Behçet's disease: a multicentric study in the Spanish population. Arthritis Research & Therapy. 2013;15(5):R145.
68. Savey L, Resche-Rigon M, Wechsler B, Comarmond C, Piette JC, Cacoub P, Saadoun D. Ethnicity and association with disease manifestations and mortality in Behçet's disease. Orphanet Journal of Rare Diseases. 2014; 9(42):1-7.
69. Jiang Y, Wang H, Yu H, Li L, Xu D, Hou S, Kijlstra A, Yang P. Two Genetic Variations in the IRF8 region are associated with Behçet's disease in Han Chinese. Scientific Reports. 2016; 6(1).
70. Kawasaki Y, Suyama K, Yugeta E, Katayose M, Suzuki S, Sakuma H, Nemoto K, Tsukagoshi A, Nagasawa K, Hosoya M. The incidence and severity of Henoch-Schönlein purpura nephritis over a 22-year period in Fukushima Prefecture, Japan. International Urology and Nephrology. 2010; 42(4):1023–1029.
71. Hata A, Noda M, Moriwaki R, Numano F. Angiographic findings of Takayasu arteritis: New classification. International Journal of Cardiology. 1996; 54:S155–S163.
72. Espinoza J, Ai S, Matsumura I. New Insights on the Pathogenesis of Takayasu Arteritis: Revisiting the Microbial Theory. Pathogens. 2018;7(3):73.
73. Grayson P, Ponte C, Suppiah R, Robson JC, Bates Gribbons K, Judge A et al. American College of Rheumatology-EULAR Classification Criteria for Takayasu's Arteritis. Arthritis and Rheumatology. 2022;7:2-23.
74. Borchers AT, Gershwin ME. Giant cell arteritis: A review of classification, pathophysiology, geoepidemiology, and treatment. Autoimmunity Reviews. 2012;11(6–7):A544–A554.
75. Ninan J, Lester S, Hill C. Giant cell arteritis. Best Practice & Research Clinical Rheumatology. 2016;30(1):169–188.
76. González-Gay MÁ, Ortego-Jurado M, Ercole L, Ortego-Centeno N. Giant cell arteritis: is the clinical spectrum of the disease changing? BMC Geriatrics. 2019;19(1).
77. De Smit E, Clarke L, Sanfilippo PG, Merriman TR, Brown MA, Hill CL, Hewitt AW. Geo-epidemiology of temporal artery biopsy-positive giant cell arteritis in Australia and New Zealand: is there a seasonal influence? RMD Open. 2017;3(2):e000531.
78. Richards BL, March L, Gabriel SE. Epidemiology of large-vessel vasculidities. Best Practice & Research Clinical Rheumatology. 2010; 24(6):871–883.
79. Li L, Neogi T, Jick S. Mortality in Patients with Giant Cell Arteritis: A Cohort Study in UK Primary Care. Arthritis Care & Research. 2018; 70(8):1251–1256.
80. Monach PA, Merkel PA. Genetics of vasculitis. Current Opinion in Rheumatology. 2010; 22(2):157–163.
81. Dillon MJ, Eleftheriou D, Brogan PA. Medium-size-vessel vasculitis. Pediatric Nephrology. 2009; 25(9):1641–1652.
82. Bae YD, Choi HJ, Lee JC, Park JJ, Lee YJ, Lee EB, Song YW. Clinical Features of Polyarteritis Nodosa in Korea. Journal of Korean Medical Science. 2006; 21(4):591.
83. Craven A, Robson J, Ponte C, Grayson PC, Suppiah R, Judge A, Watts R, Merkel PA, Luqmani RA. ACR/EULAR-endorsed study to develop Diagnostic and Classification Criteria for Vasculitis (DCVAS). Clinical and Experimental Nephrology. 2013; 17(5):619–621.
84. Jacobs-Kosmin D. Polyarteritis Nodosa: Practice Essentials, Pathophysiology, Etiology. Polyarteritis Nodosa: Practice Essentials, Pathophysiology, Etiology. 2022.
85. Mahr A, Guillevin L, Poissonnet M, Aymé S. Prevalences of polyarteritis nodosa, microscopic polyangiitis, Wegener's granulomatosis, and Churg-Strauss syndrome in a French urban multiethnic population in 2000: A capture-recapture estimate. Arthritis Care & Research. 2004; 51(1):92–99.
86. Kanecki K, Nitsch-Osuch A, Gorynski P, Wierzba W, Tarka P, Tyszko P. Polyarteritis nodosa: decreasing incidence in Poland. Archives of Medical Science. 2019; 15(5):1308–1312.
87. Eleftheriou D, Dillon MJ, Tullus K, Marks SD, Pilkington CA, Roebuck DJ, Klein NJ, Brogan PA. Systemic Polyarteritis Nodosa in the Young: A Single-Center Experience Over Thirty-Two Years. Arthritis & Rheumatism. 2013;65(9): 2476–2485.
88. Yalçınkaya F, Özçakar ZB, Kasapçopur Ö, Öztürk A, Akar N, Bakkaloğlu A, Arısoy N, Ekim M, Özen S. Prevalence of the MEFV Gene Mutations in Childhood Polyarteritis Nodosa. The Journal of Pediatrics. 2007; 151(6):675–678.
89. Burns JC. History of the worldwide emergence of Kawasaki disease. International Journal of Rheumatic Diseases. 2017; 21(1):13–15.
90. AL-Ammouri I, Al-Wahsh S, Khuri-Bulos N. Kawasaki disease in Jordan: demographics, presentation, and outcome. Cardiology in the Young. 2011; 22(4):390–395.
91. Guo MM-H, Tseng W-N, Ko C-H, Pan H-M, Hsieh K-S, Kuo H-C. Th17- and Treg-related cytokine and mRNA expression are associated with acute and resolving Kawasaki disease. Allergy. 2015; 70(3):310–318.
92. Chahal N, Somji Z, Manlhiot C, Clarizia NA, Ashley J, Yeung RSM, McCrindle BW. Rate, associated factors, and outcomes of recurrence of Kawasaki disease in Ontario, Canada. Pediatrics International. 2012; 54(3):383–387.
93. Ball G. The History of ANCA-associated Vasculitis. Rheumatic Disease Clinics of North America. 2010; 36(3):439-446.
94. WATTS RA. Epidemiology of vasculitis in Europe. Annals of the Rheumatic Diseases. 2001; 60(12): 1156a–11157.
95. Watts RA, Mahr A, Mohammad AJ, Gatenby P, Basu N, Flores-Suárez LF. Classification, epidemiology, and clinical subgrouping of antineutrophil cytoplasmic

antibody (ANCA)-associated vasculitis. Nephrology Dialysis Transplantation. 2015; 30(suppl_1):i14–i22.

96. Mohammad AJ. An update on the epidemiology of ANCA-associated vasculitis. Rheumatology. 2020; 59(Supplement_3):iii42–iii50.
97. Sugiyama K, Sada K, Kurosawa M, Wada J, Makino H. Current status of the treatment of microscopic polyangiitis and granulomatosis with polyangiitis in Japan. Clinical and Experimental Nephrology. 2012; 17(1):51–58.
98. Ugarte-Gil M, Espinoza L. 2014. Genetics of ANCA-associated Vasculitides. Current Rheumatology Reports. 2014;16(7):428.
99. Panupattanapong S, Stwalley DL, White AJ, Olsen MA, French AR, Hartman ME. Epidemiology and Outcomes of Granulomatosis with Polyangiitis in Pediatric and Working-Age Adult Populations In the United States. Arthritis & Rheumatology. 2018; 70(12):2067–2076.
100. Oni L, Sampath S. Childhood IgA Vasculitis (Henoch Schonlein Purpura)—Advances and Knowledge Gaps. Frontiers in Pediatrics. 2019;7.
101. Yagnik P, Jain A, Amponsah JK, Bhatt P, Parmar N, Donda K, Sharma M, Dave M, Chaudhari R, Vasylyeva TL, Dapaah-Siakwan F. National Trends in the Epidemiology and Resource Use for Henoch-Schönlein Purpura (IgA Vasculitis) Hospitalizations in the United States From 2006 to 2014. Hospital Pediatrics. 2019; 9(11):888–896.
102. López-Mejías R, Castañeda S, Genre F, Remuzgo-Martínez S, Carmona FD, Llorca J, Blanco R, Martín J, González-Gay MA. Genetics of immunoglobulin-A vasculitis (Henoch-Schönlein purpura): An updated review. Autoimmunity Reviews. 2018; 17(3):301–315.
103. McAdoo SP, Pusey CD. Anti-Glomerular Basement Membrane Disease. Clinical Journal of the American Society of Nephrology. 2017; 12(7):1162–1172.
104. Desbois AC, Cacoub P, Saadoun D. Cryoglobulinemia: An update in 2019. Joint Bone Spine. 2019;86(6):707–713.
105. Roccatello D, Saadoun D, Ramos-Casals M, Tzioufas AG, Fervenza FC, Cacoub P, Zignego AL, Ferri C. Cryoglobulinaemia. Nature Reviews Disease Primers. 2018; 4(1).
106. Zignego AL, Wojcik GL, Cacoub P, Visentini M, Casato M, Mangia A, Latanich R, Charles ED, Gragnani L, Terrier B, Piazzola V, Dustin LB, Khakoo SI, Busch MP, Lauer GM, et al. Genome-wide association study of hepatitis C virus- and cryoglobulin-related vasculitis. Genes & Immunity. 2014; 15(7):500–505.
107. Sjöwall C, Mandl T, Skattum L, Olsson M, Mohammad AJ. Epidemiology of hypocomplementaemic urticarial vasculitis (anti-C1q vasculitis). Rheumatology. 2018; 57(8):1400–1407.
108. Kirino Y, Nakajima H. Clinical and Genetic Aspects of Behçet's Disease in Japan. Internal Medicine. 2019; 58(9):1199–1207.
109. Salmaninejad A, Gowhari A, Hosseini S, Aslani S, Yousefi M, Bahrami T, Ebrahimi M, Nesaei A, Zal M. Genetics and immunodysfunction underlying Behçet's disease and immunomodulant treatment approaches. Journal of Immunotoxicology. 2017; 14(1):137–151.
110. Bonaguri C, Orsoni J, Russo A, Rubino P, Bacciu S, Lippi G, Melegari A, Zavota L, Ghirardini S, Mora P. Cogan's syndrome: anti-Hsp70 antibodies are a serological marker in the typical form. Isr Med Assoc J. 2014; 16(5):285-288.
111. Cogan's Syndrome - Vasculitis Foundation. Vasculitis Foundation. 2022. https://www.vasculitisfoundation.org/education/forms/cogans-syndrome/[Accessed 29 October 2022].
112. Iliescu DA, Timaru CM, Batras M, De Simone A, Stefan C. COGAN'S SYNDROME. Rom J Ophthalmol. 2015; 59(1):6-13.
113. Atisha-Fregoso Y, Hinojosa-Azaola A, Alcocer-Varela J. Localized, single-organ vasculitis: clinical presentation and management. Clinical Rheumatology. 2013; 32:1-6.