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#### **Original Article**

# Scientific Knowledge of Wegener's Granulomatosis: A Scientometric Analysis, from 1970 to 2023

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# ARTICLE INFO ABSTRACT

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#### Key words:

Granulomatosis with polyangiitis; Wegener's granulomatosis; Scientometric; Bibliometric **Introduction:** Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis is a systemic, necrotizing vasculitis. To our knowledge, there have been no previous attempts to assess the literature on GPA through scientometric analysis. In our study, we utilized scientometric analysis to explore the geographical, institutional, publication, authorship, citation, and keyword dimensions of GPA research, with the goal of uncovering the current state and emerging trends in the field.

**Methods:** The bibliographic information for studies on GPA was obtained from Scopus up to 2024. VOSviewer software was used to analyze publication characteristics, including countries, institutions, journals, authors, core references, and keywords.

**Results:** The literature review yielded 15092 publications in the Title-abstract-keyword fields related to GPA. The number of published articles increased from 2014 to 2021, and decreased since 2021. The United States (n=3672, 24.3%), has the highest publication number. There was a strong and significant positive correlation between the number of articles produced by countries on GPA and their gross domestic product (GDP) (r = 0.7103, P < 0.001). Mayo Clinic (n=353) is the most active institution and the Journal of Rheumatology (n=248) is the most active journal. The analysis of the co-occurrences of keywords was performed by VOSviewer. The most frequent author keyword was "Wegener's Granulomatosis" (n=1718).

**Conclusion:** The current study comprehensively reviewed GPA research from 1970 to 2024 using Scopusindexed articles. Results highlighted leading countries, institutions, journals, influential publications, and key authors, identifying impactful research avenues. This scientometric review offers valuable insights for future research directions and publishing strategies in GPA. By recognizing trends and emerging themes, clinicians can enhance their practice, engage in relevant research, and contribute to improved patient outcomes.

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#### Introduction

Granulomatosis with polyangiitis (GPA), initially identified as Wegener's granulomatosis, seen significant advancements has in diagnosis, treatment. and understanding of its pathophysiology, especially after the of antineutrophil discovery cytoplasmic antibodies (ANCA) in the 1980s. This disease is a systemic, necrotizing vasculitis typically associated with ANCA that target proteinase 3 (PR3) and produce a cytoplasmic staining pattern. The primary features of GPA include granulomatous and necrotizing inflammation, principally occurring in the upper and lower respiratory tracts, often accompanied rapidly progressing pauci-immune by glomerulonephritis.<sup>1, 2</sup> It is particularly more common in the Nordic countries in Europe. with an estimated annual incidence of around 10 cases per million people in northern Europe.<sup>3</sup> The precise origin of GPA remains unclear and is likely multifactorial. Environmental triggers, such as inhaling dust or exposure to silica, have been implicated in about 10% of GPA cases. There are theories suggesting that infectious agents could initiate the disease through a process known as molecular mimicry, with the nasal presence of Staphylococcus aureus possibly leading to disease flare-ups.4 Additionally, cases of GPA occurring among siblings have been documented, hinting at a familial pattern. The influence of genetic predispositions on GPA development was supported by findings from a recent genomewide association study involving 1683 GPA patients and 489 patients with microscopic polyangiitis (MPA).<sup>5</sup>

Common systemic symptoms such as fever, fatigue, and weight loss are observed in

approximately 50% of patients, although these symptoms are not unique to the condition. Symptoms related to the ear, nose, and throat (ENT) are observed in 70 to 100% of individuals at the time of diagnosis.<sup>6</sup> Lung involvement is seen in 50 to 90% of patients, presenting with alveolar hemorrhage of differing intensities and/or parenchymal nodules, which can be solitary or multiple (typically not exceeding 10), with about half of these nodules being surgically removed.<sup>7,8</sup>

GPA is a critical condition that is almost always lethal without intervention. However, thanks to increasingly standardized treatment protocols and the introduction of new biological therapies, 90% of patients achieve remission, with a survival rate of about 80% after 10 years. The current guideline for GPA treatment, as outlined by the French National Authority for Health (HAS) in November 2007, addresses systematic necrotizing vasculitis. It involves an initial phase, the induction phase, designed to quickly achieve remission within 3 to 6 months, depending on the patient's response. This is followed by a maintenance phase aimed at solidifying remission and minimizing the risk of recurrence, which lasts between 12 to 24 months.9

To the best of our knowledge, there have been no previous attempts to assess the literature on GPA through scientometric analysis. Scientometrics investigates the attributes and metrics of documented systems, using statistical methods to explore the distribution, relationships, changes, and growth of scientific publications within a certain domain.<sup>10-12</sup> Tools like VOSviewer are commonly used for visual analysis in such studies.<sup>13, 14</sup> In our research, we utilized scientometric analysis to explore the geographical, institutional, publication, authorship, citation, and keyword dimensions of GPA research, with the goal of uncovering the current state and emerging trends in the field.

# Method

The bibliographic information for studies on GPA was obtained from Scopus, a leading database for academic publications.<sup>10</sup> The dataset searches for this paper were recently updated on April 31, 2024. To gather the GPA reference database, we conducted searches of titles, abstracts, and keywords in scientific papers, employing search terms outlined in the search strategy (see appendix A), yielding a total of 15092 entries, all of which were included in this study (e.g. original articles, reviews, case reports, etc.).

Scientometric methods were then applied to extract and analyze the bibliographic data from this database. The analyzed characteristics included the names of authors, titles, publication years, source titles, volumes, issues, page counts, affiliations of authors, abstracts, keywords chosen by authors, indexed keywords, reference lists, document types, and other relevant attributes.<sup>15</sup>

Descriptive statistical analyses and the creation of graphs were performed using Microsoft Office Excel 2019, provided by Microsoft Corporation in Redmond, Washington, USA. To deeply explore the advancements across various fields of study, we adopted scientometric techniques. Additionally, we utilized VOSviewer software (Version 1.6.16) for fundamental statistical analyses and to create standard graphs.<sup>11</sup> This software is particularly useful for visualizing connections, enabling the depiction of complex relationships between authors, journals, collaborating countries, and co-citations.<sup>12</sup> Moreover, VOSviewer employs text mining to identify noun phrases within the abstracts and titles of studies, using them to generate networks, clusters, and heatmaps.<sup>11</sup> In this study, VOSviewer software was used to map the country/region collaboration network, author collaboration network along with co-citation network, journals co-citation network, and co-occurrence network of keywords.<sup>16</sup>

To resolve any issues in attributing the country of origin for publications with authors from multiple countries, we decided to designate the country of the corresponding author as the article's country of origin. This approach has been utilized in similar studies.<sup>17,18</sup> The average citation count was determined by dividing the total number of citations by the total number of articles, and this metric was recorded for each principal contributing country. Typically, the number of articles reflects the productivity of institutions, while the citation count acts as an indicator of the research's impact.<sup>19</sup> The Hirsch index (H-index) is broadly accepted as a reliable measure of scientific quality.<sup>20</sup> The impact factor (IF), obtained from the most recent Journal Citation Report (JCR), is considered as the primary indicator of a journal's prestige.<sup>21</sup> Pearson's correlation analysis was employed to evaluate the correlation strength among continuous variables, with a P value of less than 0.05 considered to be statistically significant.

In overlay visualizations, nodes are typically color-coded to represent a specific temporal variable, such as year. For example, warmer colors (like yellow or red) are commonly used to indicate more recent years, while cooler colors (like blue or green) are assigned to earlier years. This color-coding helps visualize trends over time, providing insights into the evolution of collaborations or research activities within a specific period.<sup>15</sup> The Flourish website was used to create choropleth maps.<sup>22</sup>

#### Results

# Overview of the included publications

The literature review in the Scopus database yielded 15092 publications in the Title-abstractkeyword fields related to GPA. The most popular subject areas of these publications were medicine (n=14479, 95.94%), Immunology and Microbiology (n=2035, 13.5%), and Biochemistry, Genetics and Molecular Biology (n=963, 6.4%) (Figure 1A). The most frequent article types were original articles (9939, 65.8%), review articles (2541, 16.8%), and letters (n=1108, 7.3%). The remaining publications were other types of publications (Notes, editorials, book chapters, conference papers, Erratum, and Short surveys) (Figure 1B). The quantitative analysis of published articles about GPA is shown in Figure 1C. In 2023, 516 articles were published, followed by 530 in 2022, and 663 in 2021. We can see that the number of published articles increased from 2014 to 2021, and the increase was fastest from 2019 to 2021. However, the number of published articles decreased since 2021. The top 5 most frequent languages were English (n=12086, 80.1%), German (n=745, 4.9%), French (n=672, 4.4%), Japanese (n=403, 2.7%), and Spanish (n=385, 2.5%).

# **Active countries**

The top 10 countries with the highest number of articles were the United States (n=3672, 24.3%), the United Kingdom (n=1565, 10.4%),

Germany (n=1503, 9.9%), France (n=1170, 7.8%), Japan (n=1038, 6.9%), Italy (n=785, 5.2%), The Netherlands (n=588, n=3.9%), Spain (n=460, 3.0%), Canada (n=433, 2.9%), and India (n=388, 2.6%) (Table 1). In addition, the USA's total number of citations (146713) and H-index (163) both ranked first.

The country collaboration network is shown in Figure 2A, which was generated by VOSviewer. In total, 59 countries that published more than 10 articles were included. The thickness of the line demonstrates the strength of cooperation among countries (named as total link strength, TLS). The top five TLSs were the United States, the United Kingdom, Germany, Italy, and France. The clustering analysis resulted in Six different clusters representing international. Furthermore, the overlay visualization of countries, which shows the relationship of countries and time, is shown in Figure 2B. The international collaboration density map, based on these scores is shown in Figure 2C.

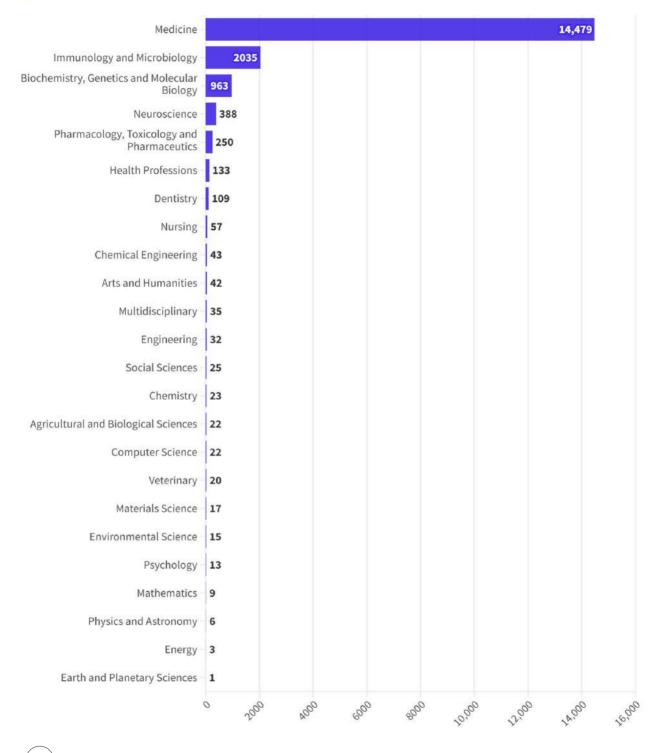
# **Correlation analysis**

There was a strong and significant positive correlation between the number of articles produced by countries on GPA and their gross domestic product (GDP) (r = 0.7103, P < 0.001). However, there was no significant correlation between the number of articles produced by countries on GPA and their human development index (HDI) or GDP per capita (P = 0.556, P = 0.239; respectively).

# Active authors

The top 10 most active authors on GPA were Gross, W.L. (n=316), Guillevin, L. (n=264), Specks, U. (n=198), Kallenberg, C.G.M.

Population 2021



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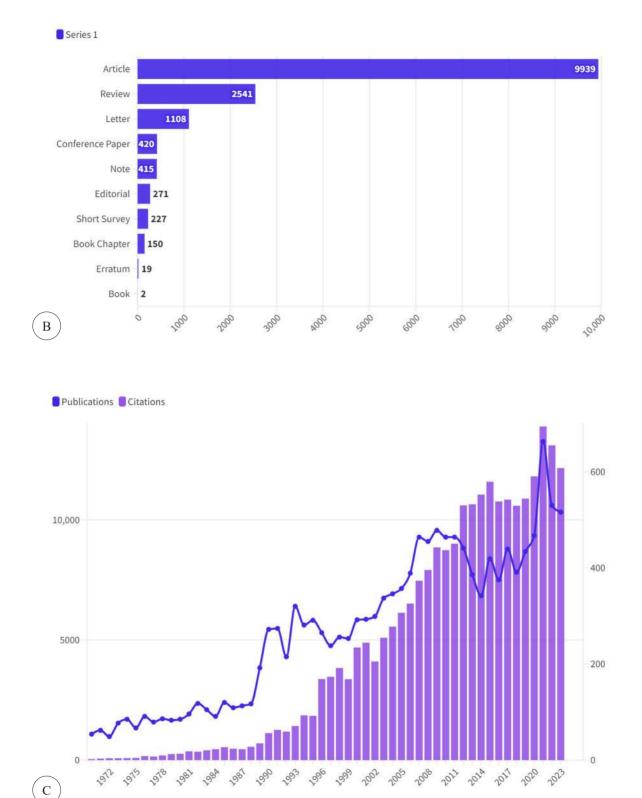
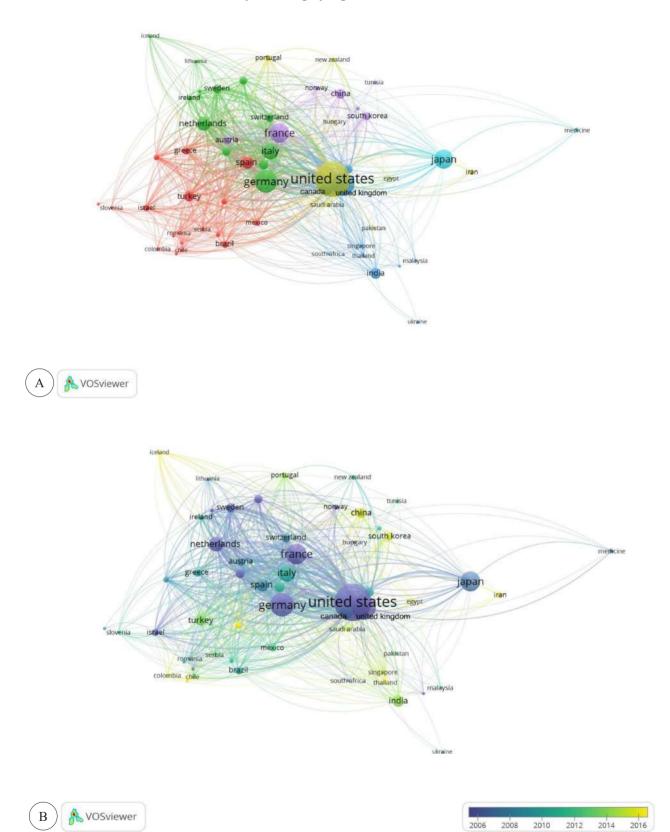


Figure 1. Number of publications in each (A) Subject area; (B) Publication type; (C) Year. Number of citations were also depicted in each year (1C).



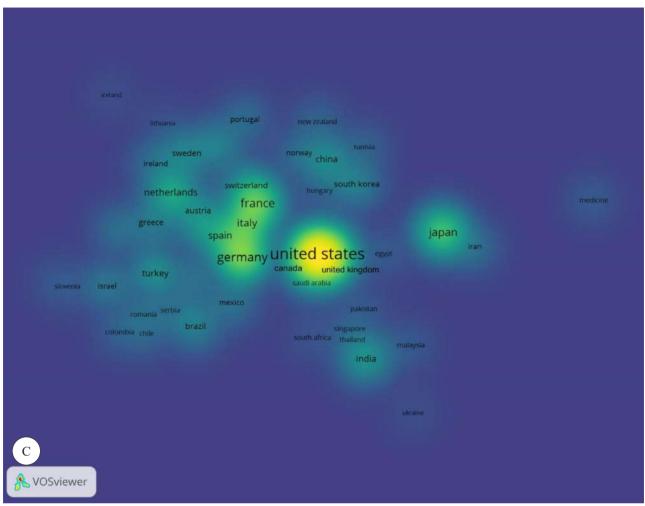


Figure 2. (A) The country collaboration network. In total, 42 countries that published more than 5 articles were included. The thickness of the line demonstrates the strength of cooperation among countries. (B) the overlay visualization of countries. (C) The international collaboration density map.

Table 1. Top 10 productive countries/regions associated with Wegener granulomatosis

rank	country	documents	percentage	TC	AAC	H-index
1	United States	3672	24.3%	146713	39.95	163
2	United Kingdom	1565	10.4%	64939	41.49	109
3	Germany	1503	9.9%	59297	39.45	110
4	France	1170	7.7%	39948	34.14	94
5	Japan	1038	6.9%	18607	17.92	53
6	Italy	785	5.2%	33488	42.66	77
7	Netherlands	588	3.9%	50314	85.57	95
8	Spain	460	3.0%	17256	37.51	50
9	Canada	433	2.9%	15075	34.81	63
10	India	388	2.6%	3667	9.45	25

TC, Total citations; AAC, Average article citations

(n=183), Hoffman, G.S. (n=148), Pagnoux, C. (n=140), Merkel, P.A. (n=124), Csernok, E. (n=120), Lamprecht, P. (n=115), and Langford, C.A. (n=115). Gross, W.L. was the most cited author with a total number of 26325 citations and had the highest H-index among the authors. However, Hoffman, G.S. average article citations (AAC) ranked first (163.65) (Table 2). The author collaboration network is shown in Figures 3A and 3B, which was generated by VOSviewer. In total, 70 authors that published more than 25 articles were included. The top five TLSs were Merkel, P.A., Specks, U., Hoffman, G.S., Seo, P., and Langford, C.A. The clustering analysis resulted in Seven different clusters representing international

collaboration.

# Active institutions

The top 10 institutions that produced the highest number of articles on GPA were as follows: Mayo Clinic (n=353), Cleveland Clinic Foundation (n=299), Hopital Cochin AP-HP (n=258), Université Paris Cité (n=254), Universitair Medisch Centrum Groningen (n=240), AP-HP Assistance Publique - Hopitaux de Paris (n=206), Universität zu Lübeck (n=197), Harvard Medical School (n=186), Massachusetts General Hospital (n=184), and Inserm (n=176). As shown in Table 3, among the top 10 productive institutions, Four come

Table 2. The 10 most productive authors with the highest number of documents.

	- 1	8				
rank	author	country	documents	TC	AAC	H-index*
1	Gross, W.L.	Germany	316	26325	83.31	76
2	Guillevin, L.	France	264	18636	70.59	60
3	Specks, U.	United States	198	18825	95.08	59
4	Kallenberg, C.G.M.	Netherlands	183	24318	132.88	62
5	Hoffman, G.S.	United States	148	24220	163.65	57
6	Pagnoux, C.	Canada	140	6139	43.85	37
7	Merkel, P.A.	United States	124	15811	127.51	52
8	Csernok, E.	Germany	120	7631	63.59	49
9	Lamprecht, P.	Germany	115	8227	71.54	32
10	Langford, C.A.	United States	115	11629	101.12	42
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TC, Total citations; AAC, Average article citations; \*, H-index only for GPA articles; The real H-index of these authors are higher

Table 3. Top 10 productive institutions in publications related to the research on Wegener granulomatosis

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rank	Institution	country	documents	TC	AAC	H-index
1	Mayo Clinic	United States	353	25076	71.04	71
2	Cleveland Clinic Foundation	United States	299	25656	85.81	62
3	Hopital Cochin AP-HP	France	258	18196	70.53	57
4	Université Paris Cité	France	254	17193	67.69	58
5	Universitair Medisch Centrum Groningen	Netherlands	240	26957	113.32	71
6	AP-HP Assistance Publique - Hopitaux de Paris	France	206	11907	57.80	50
7	Universität zu Lübeck	Germany	197	21320	108.22	60
8	Harvard Medical School	United States	186	9171	49.30	39
9	Massachusetts General Hospital	United States	184	14919	81.08	47
10	Inserm	France	176	8031	45.63	44

TC, Total citations; AAC, Average article citations

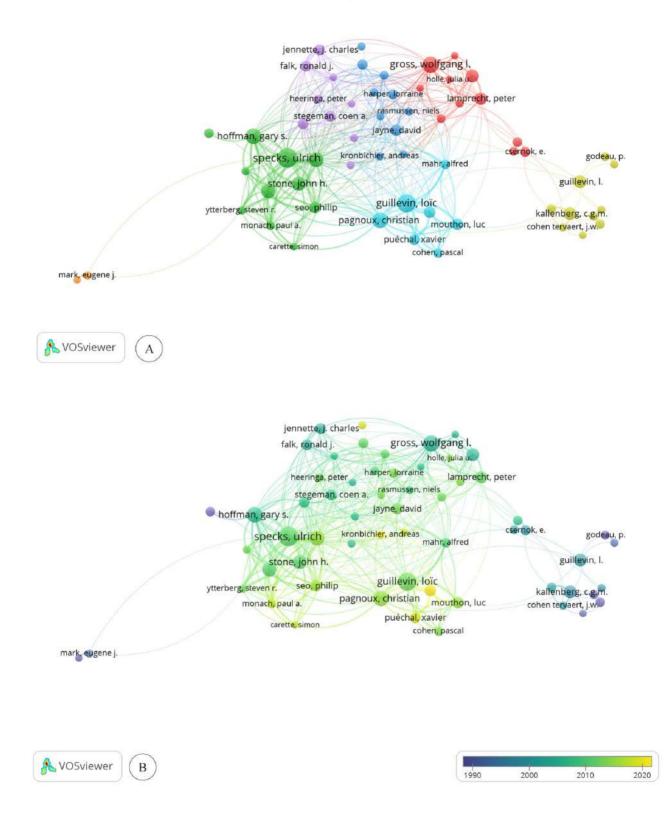


Figure 3. (A) the overlay visualization of author collaboration. (B) the network visualization of author collaboration

from the USA and Four are from the France.

#### Active journals

The top 10 journals that published the highest number of articles were Journal Of Rheumatology (n=248), Clinical And Experimental Rheumatology (n=210), Annals Of The Rheumatic Diseases (n=200), Clinical Rheumatology (n=148), New England Journal Of Medicine (n=145), Nephrology Dialysis Transplantation (n=143), BMJ Case Reports (n=141), Arthritis And Rheumatism (n=139), and Rheumatology (N=131) (Table 4).

Among the top 10 journals, 70% were in JCR Q1, and their impact factors were relatively high, with 60% being higher than 5. Besides this Arthritis And Rheumatism had the highest H-index and total citations.

Co-cited journals were analyzed by VOSviewer. The threshold was set as 700 cited times, and 78 journals were included. The top five TLSs were the Arthritis And Rheumatism, Annals Of The Rheumatic Diseases, New England Journal Of Medicine, Journal Of Rheumatology, and Annals of Internal Medicine (Figure 4A)

# **Citation analysis**

Of the 15092 articles on GPA the top 10 articles with the highest number of total citations are presented in Table 5. Interestingly, these top co-cited references were published in journals with a high impact factor, which indicates that they are authoritative articles in the research field of GPA.

In VOSviewer, the concept of "co-cited references" involves analyzing references that are cited together by other publications. Co-citation specifically occurs when two references, such as journal articles, books, or any cited works, are both cited within the same

rank	Journal	country	IF(2023)	JCR(2023)	documents	TC	AAC	H-index
1	Journal Of Rheumatology	Canada	4.66	Q1	248	6661		42
2	Clinical And Experimental Rheu- matology	Italy	4.86	Q2	210	4019		34
3	Annals Of The Rheumatic Diseases	United Kingdom	27.4	Q1	200	14459		63
4	Rheumatology	United Kingdom	5.55	Q1	162	2746		31
5	Clinical Rheumatology	United Kingdom	3.4	Q2	148	2359		25
6	New England Journal Of Medicine	United States	158.5	Q1	145	10435		20
7	Nephrology Dialysis Transplanta- tion	United Kingdom	7.186	Q1	143	4405		39
8	BMJ Case Reports	United Kingdom		Q3	141	454		10
9	Arthritis And Rheumatism	United Kingdom	10.995	Q1	139	20452		64
10	Rheumatology	United Kingdom	6.2	Q1	131	6138		46

Table 4. The 10 most productive Journals with the highest number of documents.

TC, Total citations; AAC, Average article citations; IF, IMPACT factor

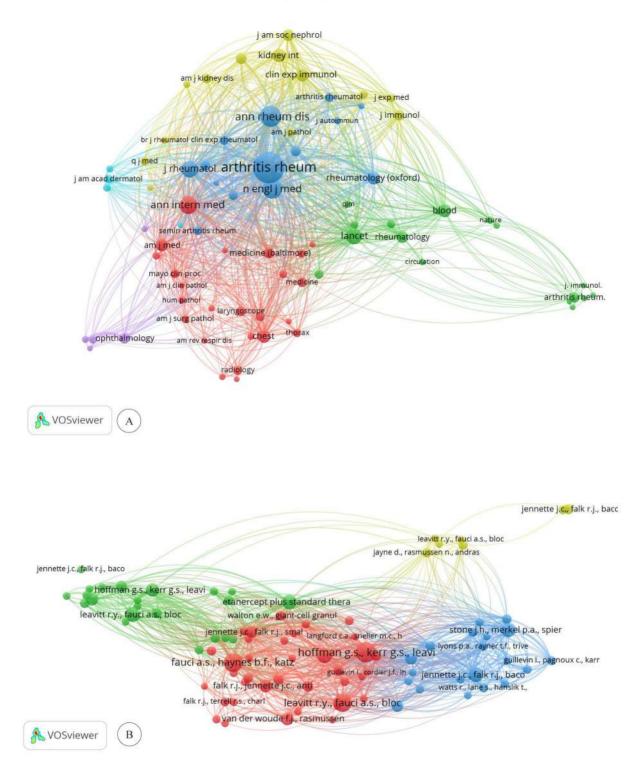


Figure 4. (A) journal-cocitation network visualization. (B) coreferences-cocitation network visualization

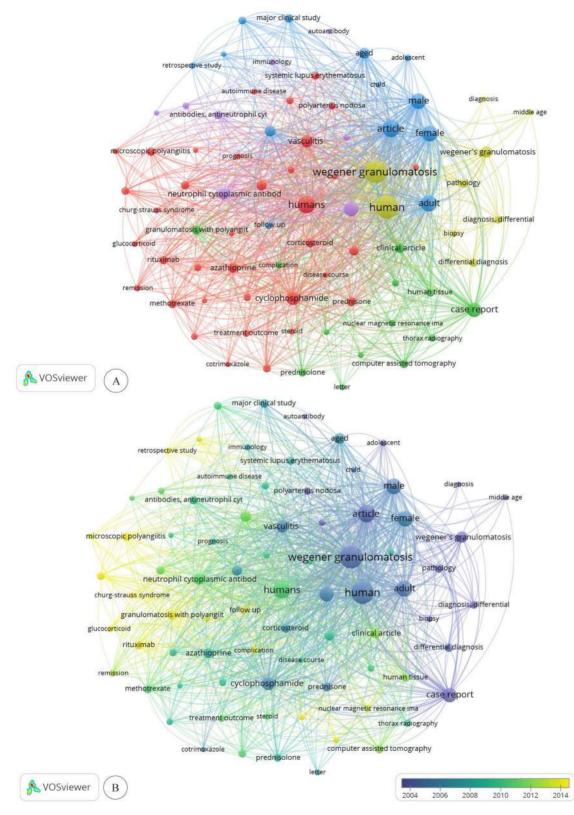


Figure 5. (A) Frequent words- cooccurrence network visualization. (B) Frequent words- cooccurrence overlay visualization

publication. The frequency with which two references are cited together across multiple publications determines the strength of their cocitation link. We used VOSviewer to analyze the co-cited references. The threshold was set as 50 cited times, and 88 references were included. The co-cited collaboration network is shown in Figure 4B.

#### **Trend topics**

The analysis of the co-occurrences of keywords was performed by VOSviewer. The most frequent words in GPA articles were Human (n=13861), Wegener Granulomatosis (n=13651), Article (n=8229), Humans (n=7981), and Adult (n=7001). Among these keywords, 79 were used in at least 800 different articles. Figures 5A and 5B displays the cluster network visualization map illustrating the

results of the cluster analysis among these keywords.

A total of 10650 author keywords were identified, and 66 keyword co-occurrences were found more than 50 times; their relationship network is shown in Figure 6A. The top five author keywords were "Wegener's Granulomatosis" (n=1718), "Vasculitis" (n=1567), "Granulomatosis with polyangiitis" (n=915), ANCA (n=603), and microscopic polyangiitis (n=423) (table 6). The network displays seven colors, representing seven clusters, and the thickness of lines reflects the relationship between keywords. The keyword "Vasculitis", with a TLS of 2539, is located in the central position of the green cluster. Furthermore, the overlay visualization of keywords, which shows the relationship of keywords and time, is shown in Figure 6B.

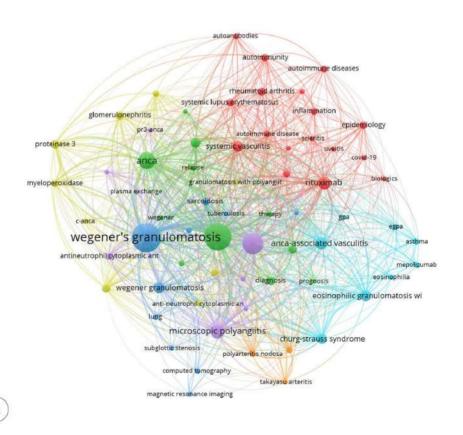
Table 5. Top 10 co-cited references concerning the research of Wegener granulomatosis

Rank	Title	Journal	Author	Year	Citations
1	2012 Revised International Chapel Hill consensus conference nomenclature of vasculitides	Arthritis and Rheumatism	Jenette, J.C.	2013	4669
2	Nomenclature of Systemic Vasculitides	Arthritis & Rheuma- tism	Jenette, J.C.	1994	3811
3	Wegener granulomatosis: An analysis of 158 patients	Annals of Internal Medicine	Hoffman, G.S.	1992	2668
4	Rituximab versus cyclophosphamide for ANCA-associated vasculitis	New England Journal of Medicine	Stone, J.H.	2010	2108
5	Nasal carriage of Staphylococcus aureus: Epidemiology, underly- ing mechanisms, and associated risks	Clinical microbiology reviews	Kluytmans, J.	1997	1968
6	Wegener's granulomatosis: Prospective clinical and therapeutic experience with 85 patients for 21 years	Annals of Internal Medicine	Fauci A.S.	1983	1748
7	The American College of Rheumatology 1990 criteria for the classification of Wegener's granulomatosis	Arthritis & Rheuma- tism	Leavitt, R.Y.	1990	1736
8	TNF-mediated inflammatory disease	Journal of Pathology	Bradley, J.R.	2008	1487
9	Autoantibodies Against Neutrophils and Monocytes: Tool For Diagnosis and Marker of Disease Activity in Wegener's Granulo- matosis	The Lancet	Van der Woude, F.J.	1985	1454
10	Anti-Neutrophil Cytoplasmic Autoantibodies with Specificity for Myeloperoxidase in Patients with Systemic Vasculitis and Idio- pathic Necrotizing and Crescentic Glomerulonephritis	New England Journal of Medicine	Falk, R.J	1988	1310

Table 6. Top 20 author keywords with the most occurrences in the included articl	es.
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Keyword	Occurrences	TLS	Keyword	Occurrences	TLS
Wegener's Granulomatosis	1718	3941	Cyclophosphamide	206	695
Vasculitis	1567	4684	Churg-Strauss syndrome	205	774
Granulomatosis with polyangiitis	915	2604	Proteinase 3	164	542
ANCA	603	2013	Antineutrophil cytoplasmic antibodies	150	469
Microscopic polyangiitis	423	1725	Rheumatoid arthritis	144	505
Rituximab	362	1221	Glomerulonephritis	143	478
ANCA-associated vasculitis	331	1019	Treatment	143	435
Wegener Granulomatosis	274	643	Diagnosis	135	346
Eosinophilic granulomatosis with polyangiitis	270	937	Systemic Lupus erythematosus	129	513
Systemic vasculitis	244	664	epidemiology	128	377

TLS, Total link strength





Scientific Knowledge of Wegener's Granulomatosis ...

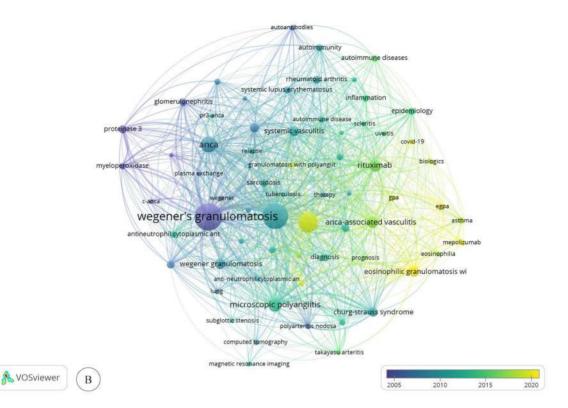


Figure 6. (A) Author keywords-cooccurrence network visualization. (B) Author keywords-cooccurrence overlay visualization

#### Discussion

To the best of our understanding, this study represents the first bibliometric analysis focused on GPA. It assesses the current state of research and serves as a valuable guide for experts in the field. Data sourced from the Scopus database as of April 31, 2023, reveals that there have been 15,092 publications related to GPA. The number of published works serves as a crucial indicator of trends within this research area.<sup>13</sup> From 2014 to 2021, the yearly output of publications has seen a steady rise, increasing from 342 to 663 articles, with the most significant growth occurring between 2019 and 2021 (Figure 1C).

With respect to countries, the United States is at the forefront of research in this specific area. In terms of geographical contributions, the United States leads in research output on this topic. The United States reports an annual incidence rate of GPA 3 per 100,000 individuals, with approximately 2,300 new cases each year. Diagnosis typically occurs between the ages of 40 and 55, with no significant difference between genders. While GPA is more common among white individuals, African Americans are somewhat underrepresented in most research, accounting for about 2-8% compared to their 11-14% representation in the overall U.S. population studied.<sup>23</sup>

A significant amount of research on GPA has been conducted in the United States, accounting for 24.3% of the global total. The US has produced 3,672 articles on the subject, while the UK follows with 1,565 articles. Additionally, the US achieved an H-index of 163 and a total citation count of 146,713. Japan ranking first in Asia and fifth globally in the number of publications, has shown notable progress in this field in recent years (Table 1). However, Japan's H-index of 53 and AAC of 17.92 are lower compared to the US and European nations, indicating the need for further improvement in research quality. Among the top 10 countries contributing to this research, only Japan and India are from Asia, and their research garners relatively fewer citations, a fact that stands out given that over 60% of the world's population resides in Asia.<sup>24</sup>

Subsequently, an examination of productive authors revealed the key figures in GPA research, with the majority based in the USA and Europe, highlighting a distinct regional influence in this field. Within the top ten highoutput authors, Four are from the United States, and 30% are from Germany. W.L. Gross from Germany stood out as the most published author and achieved the highest H-index. However, G.S. Hoffman distinguished himself with a substantially higher AAC in comparison to other authors (Table 2).

Furthermore, half of the top 10 institutions in terms of productivity are based in the USA, and all of the top ten productive institutions are of American or European origin. The Mayo Clinic is notable for publishing the most articles, receiving the highest number of citations (25,076), and achieving the highest H-index (71). However, the Universitair Medisch Centrum Groningen leads in terms of AAC (113.32). (Table 3).

The Journal of Rheumatology has been the most prolific publisher of articles related to GPA in recent years, while Arthritis and Rheumatism has received the most citations in the GPA research domain (Table 4). Considering the high quality of papers in this field, it is advised for researchers focused on GPA aim to publish their work in these journals in order to reach a wider audience.

Examining the top 10 most co-cited references, which serve as a measure of their significance through citation frequency, offers researchers insight into the key accomplishments within a specific area. Information on these references is listed in Table 5. Jenette, J.C., 2013, Arthritis and Rheumatism<sup>25</sup> and Jenette, J.C., 1994, Arthritis & amp; Rheumatism<sup>26</sup> emerged as the most cited articles, significantly influencing GPA research. It is strongly advised that clinicians and researchers focusing on this subject read these studies.

It is crucial to note that a high citation count does not necessarily indicate the high quality of a paper. A paper may be quite old and have accumulated many citations over time, or it might contain a significant limitations that leads other papers to cite it in order to highlight that issue. Therefore, one cannot conclude that a paper with more citations is inherently of higher quality. Unfortunately, no parameter has yet been developed to accurately reflect the quality of academic papers, and we are currently compelled to rely on citation counts to compare the quality of articles.

Keywords serve as a rapid method to identify the main themes and focus areas of a research study. Through an examination of the terminology employed, we can uncover the critical concepts that support the study. In our investigation, we performed an analysis of keyword co-occurrence and discovered that the terms most commonly referenced were "Wegener's Granulomatosis," "Vasculitis," "Granulomatosis with polyangiitis," ANCA, and microscopic polyangiitis (Table 6.). These keywords, mainly situated at the center of the cluster, serve as the foundational terms that highlight the main research points in this field. A similar study by Gerber et al. in 2014 critically assessed the quality and quantity of research on ANCA-associated vasculitides (AAV). He mentioned that the USA emerged as the leading contributor, accounting for 22.5% of publications and serving as a hub for international collaboration, particularly with Germany and the UK. The USA and Germany had the highest h-index and total citations, while Denmark, Sweden, and the Netherlands excelled in citation rates. In contrast, countries like Japan, China, and Turkey had limited international collaborations.<sup>27</sup>

This article provides an in-depth and cutting-edge compilation of information on GPA, aiming to comprehensively explore developments in this area comprehensively and contribute valuable insights to the existing body of knowledge. Employing VOSviewer, a tool widely used in scientometrics, this research analyzes data to enhance understanding. VOSviewer offers a variety of visualization options such as network, density, and overlay visualizations, allowing for the multifaceted presentation and analysis of bibliometric data from different perspectives. The study conducts an important analysis of terms through co-occurrence and timing within the research community's framework. These innovative analytical approaches provide fresh perspectives on the trends and shifts in the topic area, highlighting new focal points and potential paths for future research that shed light on the current status of GPA. The research was constrained by the exclusive use of the Scopus database, potentially resulting in the exclusion of pertinent data. In upcoming investigations, efforts will be made to expand the search strategy to encompass a broader

selection of databases, thus facilitating a thorough and exhaustive review of the current literature concerning GPA.

#### Conclusion

The current study provided a comprehensive review of the landscape of GPA research output up to the year 2024, as illustrated by original research articles indexed in the Scopus database. The results highlighted leading countries. institutions, journals, original publications, and key authors, helping to identify the most impactful research avenues. Although significant progress has been made in understanding GPA's etiology, pathogenesis, approaches to disease management, and the condition remains linked to substantial comorbidities and continues to negatively affect patients' quality of life. It is anticipated that this scientometric review will offer valuable insights for shaping future research directions and publishing strategies in the exploration of GPA. By identifying leading researchers and their relationships with one another, larger collaborative initiatives can be established to enhance the identification and treatment of this disease. The outcomes of these substantial collaborative efforts can greatly contribute to the advancement of scientific knowledge regarding GPA.

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Not applicable

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#### **Consent to participate**

Not applicable because no patients were involved in the study

#### **Consent for publication**

The authors all agree for submission and publication of the manuscript

# Availability of data and materials

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

#### **Competing interests**

The authors have no conflicts of interest to declare that are relevant to the content of this article.

#### **Authors' contributions**

Amir Kasaeian: Conceptualization, Methodology, Supervision Majid Alikhani: Conceptualization, Validation, Supervision Hediyeh Alemi: Investigation, Methodology Naghmeh Khavandgar: Investigation, Methodology, Javad Seyedhosseini: Investigation Elham Farhadi: Investigation Mahdi Mahmoudi: Investigation Majid Sorouri: Conceptualization, Validation, Supervision Iman Menbari Oskouie: Conceptualization, Methodology, Formal analysis, Writing -**Original Draft** 

Hoda Kavoosi: Conceptualization,

Methodology, Supervision

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# References

1. Schilder AM. Wegener's granulomatosis vasculitis and granuloma. Autoimmunity reviews. 2010;9(7):483-7.

2. Banerjee P, Jain A, Kumar U, Senapati S. Epidemiology and genetics of granulomatosis with polyangiitis. Rheumatology International. 2021;41(12):2069-89.

3. Gibelin A, Maldini C, Mahr A, editors. Epidemiology and etiology of wegener granulomatosis, microscopic polyangiitis, churg-strauss syndrome and goodpasture syndrome: vasculitides with frequent lung involvement. Seminars in respiratory and critical care medicine; 2011: © Thieme Medical Publishers.

4. Laudien M, Gadola SD, Podschun R, Hedderich J, Paulsen J, Reinhold-Keller E, et al. Nasal carriage of Staphylococcus aureus and endonasal activity in Wegener's granulomatosis as compared to rheumatoid arthritis and chronic rhinosinusitis with nasal polyps. Clinical & Experimental Rheumatology. 2010;28(1):S51.

5. Lyons PA, Rayner TF, Trivedi S, Holle JU, Watts RA, Jayne DR, et al. Genetically distinct subsets within ANCA-associated vasculitis. New England Journal of Medicine.

2012;367(3):214-23.

6. Trimarchi M, Sinico RA, Teggi R, Bussi M, Specks U, Meroni PL. Otorhinolaryngological manifestations in granulomatosis with polyangiitis (Wegener's). Autoimmunity reviews. 2013;12(4):501-5.

7. Kamali S, Erer B, Artim-Esen B, Gul A, Ocal L, Konice M, et al. Predictors of damage and survival in patients with Wegener's granulomatosis: analysis of 50 patients. The Journal of rheumatology. 2010;37(2):374-8.

8. Rodrigues AJ, Jacomelli M, Baldow RX, Barbas CV, Figueiredo VR. Laryngeal and tracheobronchial involvement in Wegener's granulomatosis. Revista Brasileira de Reumatologia. 2012;52:231-5.

9. Wallace ZS, Miloslavsky EM. Management of ANCA associated vasculitis. Bmj. 2020;368.

10. Mingers J, Leydesdorff L. A review of theory and practice in scientometrics. European journal of operational research. 2015;246(1):1-19.

11. Van Eck N, Waltman L. Software survey: VOSviewer, a computer program for bibliometric mapping. scientometrics. 2010;84(2):523-38.

12. Waltman L, Van Eck NJ, Noyons EC. A unified approach to mapping and clustering of bibliometric networks. Journal of informetrics. 2010;4(4):629-35.

13. Gao Y, Shi S, Ma W, Chen J, Cai Y,

Ge L, et al. Bibliometric analysis of global research on PD-1 and PD-L1 in the field of cancer. International immunopharmacology. 2019;72:374-84.

14. Siegel RL, Miller KD, Fuchs HE, JemalA. Cancer statistics, 2021. Ca Cancer J Clin.2021;71(1):7-33.

15. Oskouie IM, Alemi H, Khavandgar N, Mardani-Fard HA, AleTaha A, Mousavian A-H, et al. Global Research Trends on Colorectal Cancer (2014-2023): A Scientometric and Visualized Study. Archives of Iranian Medicine. 2024;27(10):563.

16. Alemi H, Khavandgar N, Oskouie IM, Mardani-Fard HA, Rostami T, AleTaha A, et al. Global research trends on systemic lupus erythematosus and thyroid cancers (1964– 2023): A scientometric and visualized study. Medicine. 2024;103(26):e38511.

17. Akyol A, Kocyigit BF. Publication activity in the field of Sjögren's syndrome: a ten-year Web of Science based analysis. Rheumatology International. 2021;41(4):763-9.

18. Zhao X, Chen J, Pan Y, Feng H, Meng
B, Meng Y. A bibliometric analysis of the global research in ankylosing spondyloarthritis (2008–2017). Rheumatology International. 2019;39(6):1091-7.

19. Ioannidis JP, Boyack KW, Baas J. Updated science-wide author databases of standardized citation indicators. PLoS biology. 2020;18(10):e3000918. 20. Kreiner G. The slavery of the h-index measuring the unmeasurable. Frontiers in human neuroscience. 2016;10:556.

21. Kaldas M, Michael S, Hanna J, Yousef GM. Journal impact factor: a bumpy ride in an open space. Journal of Investigative Medicine. 2020;68(1):83-7.

22. [Available from: https://flourish. studio/.

23. Hoffman GS, Kerr GS, Leavitt RY, Hallahan CW, Lebovics RS, Travis WD, et al. Wegener granulomatosis: an analysis of 158 patients. Annals of internal medicine. 1992;116(6):488-98.

24. Dou Y, Huang Q, He C, Meng S, Zhang Q. Rapid population growth throughout Asia's earthquake-prone areas: A multiscale analysis. International journal of environmental research and public health. 2018;15(9):1893.

25. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, et al., editors. 2012 Revised International Chapel Hill consensus conference nomenclature of vasculitides. Arthritis and Rheumatism; 2013.

26. Jennette JC, Falk RJ, Andrassy K, Bacon PA, Churg J, Gross WL, et al. Nomenclature of Systemic Vasculitides. Arthritis & Rheumatism. 1994;37(2):187-92.

27. Gerber A, Klingelhoefer D, Groneberg D, Bundschuh M. Antineutrophil cytoplasmic antibody-associated vasculitides: a scientometric approach visualizing worldwide research activity. Int J Rheum Dis. 2014;17(7):796-804.