

R E V I E W

Bibliometric analysis of the literature on von Willebrand disease: research status and trends

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Abstract. *Background and aim:* Von Willebrand disease (VWD) is considered the most prevalent inherited bleeding disorder. The current study aims to demonstrate the research status and trends on VWD worldwide. *Methods:* Bibliometric analysis was used to investigate the global research productivity and trends on VWD. The publications on VWD from 1956 to 2021 were extracted using the Web of Science database. In the VWD domain, a total of 3,643 records were analyzed for authorship and collaboration patterns, yearly productivity, highly cited documents, relevant source of publication, most prolific scholars, productive countries, and organizations. *Results:* The most productive journal, author, organization, and country were ‘Haemophilia’ with 439 publications, ‘Favaloro EJ’ with 119 publications, the ‘University of Milan’ with 192 publications, and the United States of America (USA) with 1,048 publications, respectively. The document with the highest citations was ‘Srivastava A, 2013, Haemophilia,’ which received 1,154 citations in total. In 2016, the highest number of publications shared by two author patterns was 28. With 199 publications, the year 2021 remained on the top, while the citation-wise analysis identified 2006 as the top year with 5,379 citations. *Conclusions:* Research productivity and publication trends on VWD revealed that the USA emerged as the most significant contributing country. The ‘University of Milan’ was the most significant contributing organization, while ‘Favaloro EJ’ was the most significant author. ‘Haemophilia’ was found to be the most significant journal in the field of VWD. It is recommended that researchers from countries with significant contributions to the field should collaborate with researchers from Asian countries and other countries that lack behind in research in the domain of VWD.(www.actabiomedica.it)

Key words: von Willebrand disorder, hemophilia, hemorrhagic disorder, inherited blood coagulation disorder, hematologic disease, coagulation protein disorder, bibliometrics

1. Introduction

Von Willebrand disease (VWD) is the most prevalent inherited bleeding disorder caused by quantitative or qualitative abnormalities of the von Willebrand factor (1, 2). It affects up to 1% of the total population and is equally prevalent in both genders, but the clinical

presentation of the condition is more common in women due to menstrual cycles and pregnancy (3). In recent years, advances are made in the understanding of the pathogenesis of VWD, but concerns remain pertaining to the disease’s definition, diagnostic dilemma when von Willebrand factor levels evolve with age and laboratory issues that make the classification of the various types of

VWD challenging (4). Moreover, in 2006, the International Society of Thrombosis and Haemostasis (ISTH) proposed the VWD classification to create a minimal set of VWD classes to aid in patient management (5). Prior to these guidelines, there were over 20 different VWD categories that lacked well-defined clinical usefulness (5). The ISTH classification has been in use for over a decade but it has some limitations, as problems still arise when classifying patients, even when performing all the recommended von Willebrand factor assays (6). In order to overcome these obstacles, it is necessary to have adequate knowledge of the current research trends.

Bibliometric analysis is a statistical technique applied to describe the research productivity and trends in a certain field (7). The purpose of a bibliometric study is to evaluate the rate of development, assess the current situation of scientific output, and identify the most influential contributions in a certain field (7,8). Moreover, bibliometric measures such as citation analysis can help identify the most significant investigators and institutions on the subject, as well as areas that require increased research attention (9). This information can assist funding agencies in efficiently guiding their resources. Besides, medical practitioners need to be updated with current research trends. Therefore, we aim to conduct a bibliometric analysis to analyze the global research productivity and trends on VWD.

2. Objectives

The primary objective of this study was to analyze the research trends and literature published on VWD from 1956 to 2021. The sub-objectives were as follows:

1. To evaluate the authorship and collaboration patterns
2. To examine the yearly productivity figures published on the topic
3. To find out the most relevant sources of publication
4. To identify the most productive authors, organizations, and countries
5. To analyze publishing and citation trends over time
6. To identify the most commonly used keywords

3. Methods

Our target database to download the bibliographic records on VWD research output was the Web of Science (WOS). WOS is one of the most widely used and authoritative databases of research publications and citations (10-12).

The data for the present bibliometric analysis were obtained from the WOS Core Collection on January 26, 2022. Data were downloaded using advanced search as well as the topic (TS) search option, using the following search query: TS= ("Hemophilia, Vascular "OR" Vascular Hemophilia "OR" Vascular Hemophilias "OR" Vascular Pseudohemophilia "OR" Pseudohemophilia, Vascular "OR" Pseudohemophilias, Vascular "OR" Vascular Pseudohemophilias "OR" Von Willebrand's Factor Deficiency "OR" Von Willebrand Disorder "OR" Disorder, Von Willebrand "OR" von Willebrand's Disease "OR" von Willebrand's Diseases "OR" angiohemophilia "OR" angiohemophilias "OR" von Willebrand Disease "OR" von Willebrand Disease "OR" Recessive Form").

A comprehensive search and selection strategy was used to incorporate all relevant literature linked to VWD using the Boolean operator "OR" without applying any filters such as time period, country, or language for the data to be downloaded. In the 66 years between 1956 and 2021, the initial search retrieved 5,453 documents. All search results were evaluated for relevance, and documents that didn't match the search criteria were instantly eliminated. The following documents (n=1,810) were eliminated from the analysis: meeting abstracts (1,244), letters (299), editorial material (162), notes (41), early access (22), corrections (19), news items (6), book chapters (2), reprints (2), corrections, additions (1), discussions (1), and (11) records for the year 2022 were omitted from the analysis. The current investigation was limited to WOS-indexed publications on "von Willebrand disease". No other databases were used for the extraction of the bibliographic records. Finally, 3,643 records were considered. Journal articles (n=2,915), conference papers (n=182), and reviews (n=546) were included in the analysis. These records were exported in order to be analyzed. The data were analyzed using MS Excel (v16.0), MS Access, VOS viewer (version 1.6.17), Biblioshiny

(version 3.1.4), and CiteSpace (version 5.8 R3) to investigate the type of documents, authorship and collaboration patterns, yearly productivity, highly cited documents, relevant source of publication, most prolific scholars, productive organizations and countries, publishing and citation trends, commonly used keywords, country and journal citation bursts to obtain the most relevant and comprehensive results.

4. Results

4.1 Type of documents

This study reports the analysis of data retrieved from 1956 to 2021. Out of 3,643 documents, the document type 'Article' appeared as the most preferred type that the researchers in this field used to share their research (Table 1). The document type 'Review' remained the second most preferred type, followed by 'Proceedings Paper'. The same pattern was observed

regarding citations secured by all the document types. The document type 'Article' secured the highest number of citations and maintained the top position in securing citations, followed by 'Review' and 'Proceedings Paper'.

4.2 Authorship patterns

Figure 1 depicts the authorship patterns in the field of VWD. The analysis reveals various authorship patterns consisting of a single author to twenty-six authors. The data present a trend of collaborative research

Table 1. Type of documents.

Documents	TP	Percentage	TC
Article	2,915	80.02	71,063
Proceedings Paper	182	5.00	5,018
Review	546	14.99	19,663
Total	3,643	100	95,744

Abbreviations: TP, total publications; TC, total citations

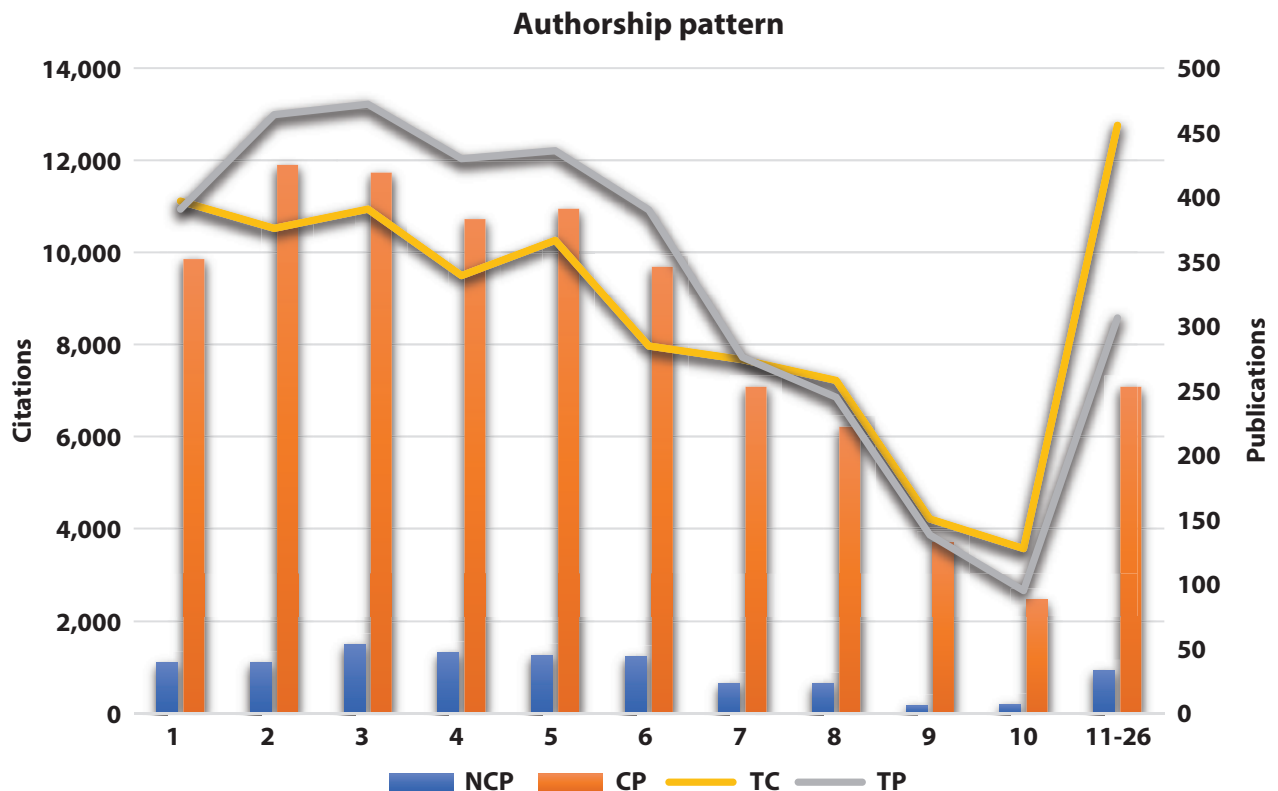


Figure 1. Authorship pattern.

in this field, with the three-author pattern at the top position by contributing 472 publications. The two-author, three-author, and five-author patterns maintained the second, third, and fourth positions. The single-author pattern attained the fifth position with 391 publications. The citation analysis presents some interesting findings. The single-author pattern, which maintained the fifth position in publishing research, secured the highest number of citations and held the first position, followed by three, two, and five-author patterns. The authorship patterns ranging from eleven

to twenty-six authors collectively secured 12,758 citations. The analysis also illustrates not cited papers (NCP) and cited papers (CP) against each authorship pattern. The three-author pattern had the highest number of not cited papers (NCP), followed by four and five-author patterns.

4.3 Authorship trend over the years

Table 2 provides the breakup of authorship patterns over the years. According to the data, the

Table 2. Authorship over the years.

PY, AU	1	2	3	4	5	6	7	8	9	10	11-26	Total
1956-1995	25	35	45	58	46	35	29	20	7	4	11	315
1996	14	15	12	15	4	16	7	7	3	2	2	97
1997	11	10	10	4	7	13	3	7	2	3	2	72
1998	17	10	17	14	12	7	10	2	4	4	2	99
1999	24	15	12	8	10	9	10	7	2		4	101
2000	15	10	6	10	18	9	11	5	3	3	1	91
2001	14	14	11	14	17	13	8	5	1	3	3	103
2002	14	8	13	20	13	5	8	8	6	3	2	100
2003	8	6	7	14	10	11	8	7	4	1	3	79
2004	10	12	11	10	13	7	8	6	4	3	4	88
2005	20	14	10	13	13	9	6	4	1	4	5	99
2006	23	15	14	14	11	18	13	10	4	2	11	135
2007	20	24	24	15	15	5	3	12	5	3	9	135
2008	17	24	21	11	20	12	10	6	9	2	10	142
2009	16	11	19	13	14	18	8	9	11	3	6	128
2010	12	21	22	12	13	12	8	9	6	3	12	130
2011	30	18	19	18	24	22	10	14	8	2	15	180
2012	15	24	25	23	20	13	6	16	4	2	9	157
2013	15	20	27	16	13	17	12	8	9	8	16	161
2014	11	13	21	17	17	16	13	11	2	3	8	132
2015	8	10	13	14	14	5	7	13	5	5	18	112
2016	9	28	18	12	11	20	12	8	5	2	25	150
2017	6	22	14	9	17	19	9	11	5	10	19	141
2018	8	22	20	23	24	26	13	8	7	3	24	178
2019	6	23	16	19	16	14	14	9	7	4	19	147
2020	15	21	21	12	19	15	12	12	9	6	30	172
2021	8	19	24	22	25	24	18	11	5	7	36	199
Total	391	464	472	430	436	390	276	245	138	95	306	3643

Abbreviations: PY, publication year; AU, number of authors

highest number of single-author publications appeared in 2011, whereas single-authors published 25 documents from the year 1956 to 1995. Likewise, 28 was the highest number of publications shared by two-author patterns in 2016, and this pattern contributed 35 documents from 1956 to 1995. Table 2 also highlights the other authorship pattern contributions over the years.

4.4 Yearly growth

As shown in Table 3, the research on VWD seems to have begun in 1956, with only two publications in this year. The analysis reveals a slow growth with single-digit published research on the topic with gaps in many years. The research on the topic started appearing without any gap in 1990. The year 1991 witnessed an instant growth in the research with 61 publications. After that, except in a few years, over 100 publications were contributed on the topic every year. The year 2021 was at the top with 199 publications, followed by 2011, 2018, and 2020. The citation-wise analysis ranked 2006 at the top with 5,379 citations, followed by 2013, 1998, and 2008. The study also describes Not Cited Papers (NCP) and Cited Papers (CP) against each year.

4.5 Most cited documents

Table 4 highlights the top 10 most cited documents on VWD. The analysis ranked the document 'Srivastava A, 2013, Haemophilia' at the top with the highest number of citations, followed by 'Hemker HC, 2003, Pathophysiol Haemost Thromb', 'Sadler JE, 1998, Annu Rev Biochem', and 'Sadler JE, 2006,

Table 3. Yearly productivity.

PY	NCP	CP	TP	TC
1956	0	2	2	89
1959	0	3	3	54
1962	0	1	1	10
1963	0	1	1	5
1964	0	1	1	47
1968	1	1	2	30

PY	NCP	CP	TP	TC
1974	0	1	1	16
1975	1	0	1	0
1978	0	2	2	39
1981	1	0	1	0
1986	1	1	2	15
1987	0	2	2	694
1990	0	4	4	58
1991	1	60	61	2604
1992	3	63	66	2124
1993	2	56	58	1998
1994	2	44	46	1421
1995	2	59	61	2553
1996	4	93	97	3538
1997	4	68	72	1861
1998	4	95	99	5024
1999	6	95	101	2820
2000	3	88	91	3877
2001	2	101	103	3629
2002	5	95	100	3731
2003	2	77	79	4679
2004	5	83	88	3384
2005	4	95	99	3623
2006	5	130	135	5379
2007	6	129	135	4563
2008	12	130	142	4986
2009	5	123	128	3207
2010	5	125	130	3914
2011	10	170	180	4451
2012	6	151	157	3168
2013	9	152	161	5304
2014	5	127	132	2744
2015	4	108	112	2392
2016	8	142	150	2463
2017	7	134	141	1895
2018	20	158	178	1433
2019	27	120	147	835
2020	52	120	172	543
2021	125	74	199	544
Total			3,643	95,744

Abbreviations: PY, publication year; NCP, not cited papers; CP, cited papers; TP, total publications; TC, total citations

Table 4. Top 10 most cited documents.

Documents (n=3,643)	Total citations	Citation years	TC per year
Srivastava A, 2013, Haemophilia	1,154	9	128.22
Hemker HC, 2003, Pathophysiol Haemost Thromb	1,035	19	54.47
Sadler JE, 1998, Annu Rev Biochem	1,010	24	42.08
Sadler JE, 2006, J Thromb Haemost	774	16	48.38
Furlan M, 1996, Blood	693	26	26.65
Gill JC, 1987, Blood	693	35	19.80
Nichols WL, 2008, Haemophilia	574	14	41.00
Kozek-Langenecker SA, 2013, Eur J Anaesth	532	9	59.11
Hemker HC, 2002, Pathophysiol Haemost Thromb	513	20	25.65
Bolton-Maggs PHB, 2003, Lancet	463	19	24.37

Abbreviations: TC, total citations

Table 5. Source of publications.

Source (n=774)	TP	TC	CP	NCP	TC/TP	h-index	PY start
Haemophilia	439	11,125	414	25	25.34	47	1995
Journal of Thrombosis and Haemostasis	208	9,257	203	5	44.50	52	2003
Thrombosis and Haemostasis	208	6,892	202	6	33.13	46	1990
Blood	165	11,399	163	2	69.08	60	1978
Seminars in Thrombosis and Hemostasis	154	4,592	152	2	29.82	36	1995
Blood Coagulation & Fibrinolysis	118	2,229	109	9	18.89	27	1991
Thrombosis Research	114	1,928	111	3	16.91	23	1978
American Journal of Hematology	72	1,886	70	2	26.19	27	1991
Hamostaseologie	50	380	41	9	7.60	11	2008
Clinical and Applied Thrombosis-Hemostasis	48	592	44	4	12.33	14	1995

Abbreviations: TP, total publications; TC, total citations; CP, cited papers; NCP, not cited papers; PY, publication year

J Thromb Haemost'. The oldest document in the list, 'Gill JC, 1987, Blood' remained at the sixth position.

4.6 Most productive sources

The top 10 most productive sources are shown in Table 5. The analysis ranked 'Haemophilia' at the top position with 439 publications. The sources 'Journal of Thrombosis and Haemostasis' and 'Thrombosis and Haemostasis' maintained the second position due to publishing an equal number of documents. The publishing source 'Blood' at the third position in publishing documents on the topic obtained the highest number of citations, followed by 'Haemophilia', 'Journal of Thrombosis and Haemostasis', and

'Thrombosis and Haemostasis'. Regarding Not Cited Papers (NCP), the source 'Haemophilia' had 25 publications that could not obtain any citation. The other prominent publishing sources 'Blood Coagulation & Fibrinolysis' and 'Hamostaseologie', each had nine publications that did not receive any citations.

4.7 Most prolific authors

Table 6 reports the top 10 prolific authors. Researcher 'Favaloro EJ' contributed 119 publications since 1991 and remained at the top, followed by 'Federici AB', and 'Castaman G'. The citation-wise analysis ranked 'Federici AB' at the top with 6664 citations from 111 publications, followed by 'Rodeghiero

F' and 'Mannucci PM'. The authors 'Federici AB' and 'Mannucci PM' jointly maintained the top position in terms of h-index, followed by 'Castaman G'.

4.8 Most productive organizations

The top 10 most productive organizations publishing research on the topic are presented in Table 7. The 'University of Milan' emerged as the most productive organization due to the highest number of publications, followed by 'Queen's University', and 'Westmead Hospital'. The citation analysis also ranked the 'University of Milan' at the top with the highest number of citations, followed by 'San Bortolo Hospital', and 'Leiden University'.

4.9 Citation pattern

The citation pattern on VWD is presented in Table 8. It is evident from the data that 359 publications did not obtain any citations. There are 235, 197, and 138 publications that received one, two, and three citations, respectively. There are 1781 publications that obtained citations between 11 to 99.

4.10 Most prolific countries with their impact

Figure 2 illustrates the research contribution from different countries and continents with their impact. There were 20 countries from Europe, nine from Asia, and three from South America that contributed to the

Table 6. Top 10 contributing authors.

Author (n=10,492)	TP	TC	PY start	h-index
Favaloro EJ	119	4394	1991	36
Federici AB	111	6664	1992	44
Castaman G	105	5173	1990	41
Budde U	103	5286	1993	38
Mannucci PM	102	5389	1991	44
Schneppenheim R	79	3696	1993	31
Rodeghiero F	73	5619	1990	38
Mazurier C	72	3772	1991	29
Lillicrap D	71	3486	1991	28
Casonato A	65	1088	1991	20

Abbreviations: TP, total publications; TC, total citations; PY, publication year

Table 7. Top 10 contributing organizations.

Organizations	NCP	CP	TP	TC
University of Milan	7	185	192	9,841
Queen's University	7	138	145	5,041
Westmead Hospital	3	106	109	4,005
Leiden University	6	99	105	5,086
Medical College of Wisconsin	9	92	101	4,497
San Bortolo Hospital	1	91	92	6,250
University of Padua	2	73	75	1,363
Royal Free Hospital	1	64	65	4,377
Lund University	5	55	60	2,787
University of Michigan	3	56	59	2,511

Abbreviations: NCP, not cited papers; CP, cited papers; TP, total publications; TC, total citations

Table 8. Citation pattern.

TC	TP
0	359
1	235
2	197
3	138
4	129
5	139
6	127
7	99
8	93
9	90
10	102
11-99	1781
100-199	106
200-1154	48

Abbreviations: TP, total publications; TC, total citations

research on VWD. The USA contributed the highest number of publications with 1048, followed by Italy, Germany, and the UK. The USA also topped in securing the highest number of citations, followed by Italy, the UK, the Netherlands, and France.

4.11 Thematic focus and trend topics

The timeline visualization of topic trends on VWD research is shown in Figure 3. The timeline from left to right presents the clusters of the documents related to VWD research, and the publication years are shown at the bottom of the graph. The figure describes the thematical evolution of the topic related to VWD research. The topics in the figure have been arranged vertically, mentioning the latest topics at the top and the oldest topics at the bottom. The topics 'heavy menstrual bleeding', 'blood coagulation disorders', and 'hemorrhage' appeared at the top of the timeline demonstrating the latest trends. On the contrary, the topics 'bleeding time', 'ddavp', 'ristocetin cofactor activity', and 'platelet function' appeared at the bottom of the timeline.

Figure 4 portrays the most frequently used author keywords. The author-keyword 'Von Willebrand Disease' emerged as the most frequently used

keyword by researchers on the topic. The author-keyword 'Von Willebrand Factor' happened to be the second most commonly used keyword, followed by 'Hemophilia' and 'Bleeding Disorder'. The figure also presents other most frequently used keywords on VWD research.

Figure 5 presents the author-keyword map generated by VOSviewer visualization software with zero minimum total link strength of an item. Out of 43 items, all met the threshold. The VOSviewer software divided all the keywords into seven clusters with 479 links and 4277 total link strengths. The software gathered all the keywords with topic similarity in one cluster. The size of the circles in the map depends on the frequency of the keywords used by the authors. The bigger size of the circle shows the higher trend of the usage of the keywords by the authors. The keywords like von Willebrand disease, von Willebrand factor, bleeding disorder, hemophilia, and desmopressin appeared as the most frequently used keywords.

4.12 Top 10 countries with the strongest citation bursts

Citation bursts play an essential role in identifying the items that have achieved rapid growth in citations in a specific period. It depicts the dynamics and direction of a research field. The analysis of items obtaining instant growth in citations helps identify trends in a particular research field that has been receiving the attention of the researchers. Figure 6 portrays the top 10 countries with their strongest citation bursts. The figure reveals that the earliest citation bursts on the topic from these countries began in 1974. The citation bursts depict that the research related to VWD from these countries got great attention from 1974 to 2021. The red line on the figure shows the range of the citation bursts for each top country.

4.13 Top 10 journals with the strongest citation bursts

Figure 7 presents the top 10 cited journals with the strongest citation bursts related to VWD research. The figure depicts the start of the citation burst in 1956. The citation bursts show the research direction of the journals on the topic from 1956 to 2021.

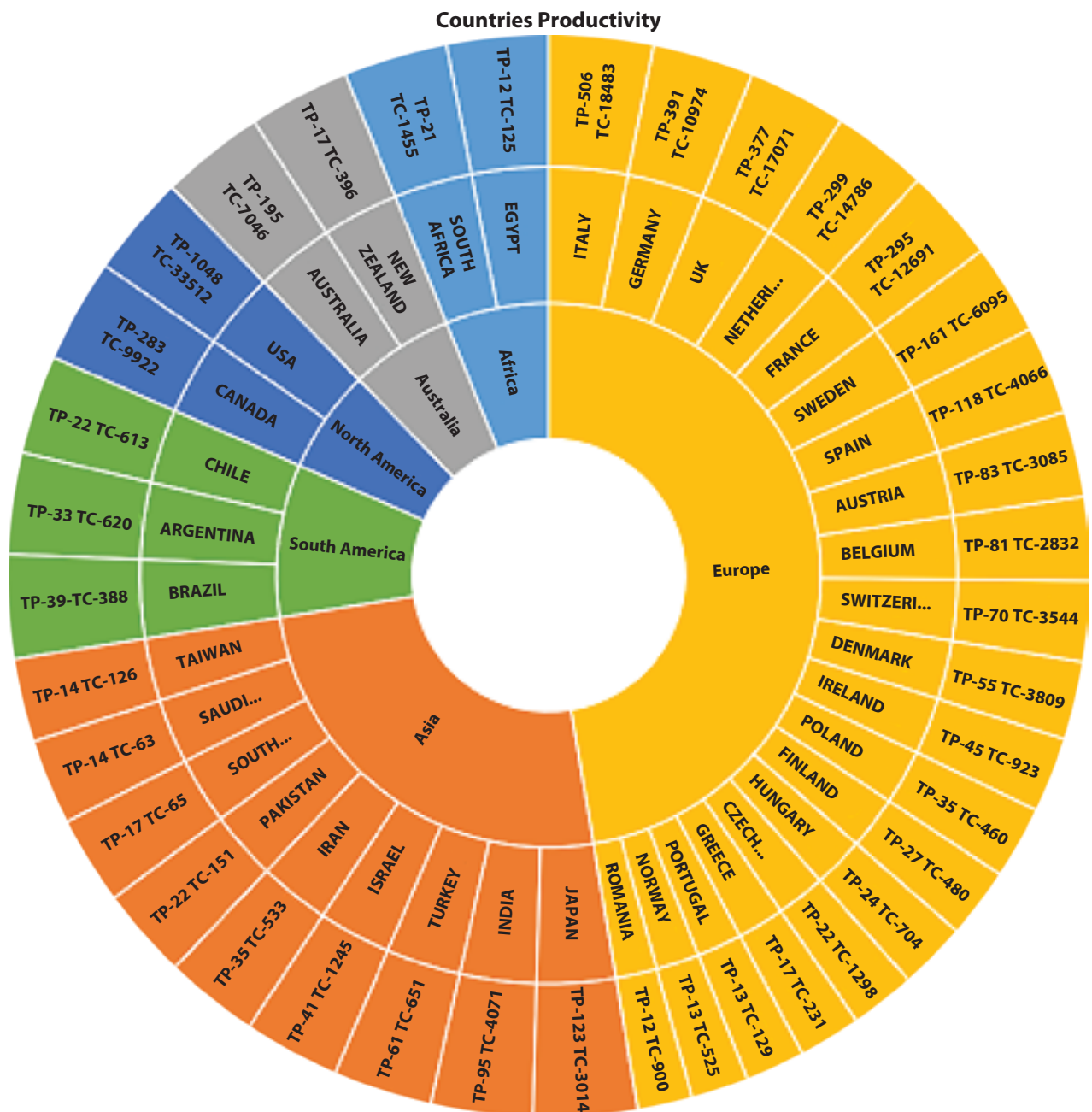


Figure 2. Country productivity.

5. Discussion

5.1 Chronological distribution of publications

According to our results, VWD was first mentioned in a journal in 1956, as determined through our review of WOS. During the period from 1956 to 1990,

the total number of publications remained steady. There was a significant increase in the total number of publications on VWD starting in 1991. In the year 1999, it was witnessed for the first time that 100 papers on VWD were published in a single year. Recent investigations have shown that in broader domains such as cardiology, and nephrology, a tendency of more

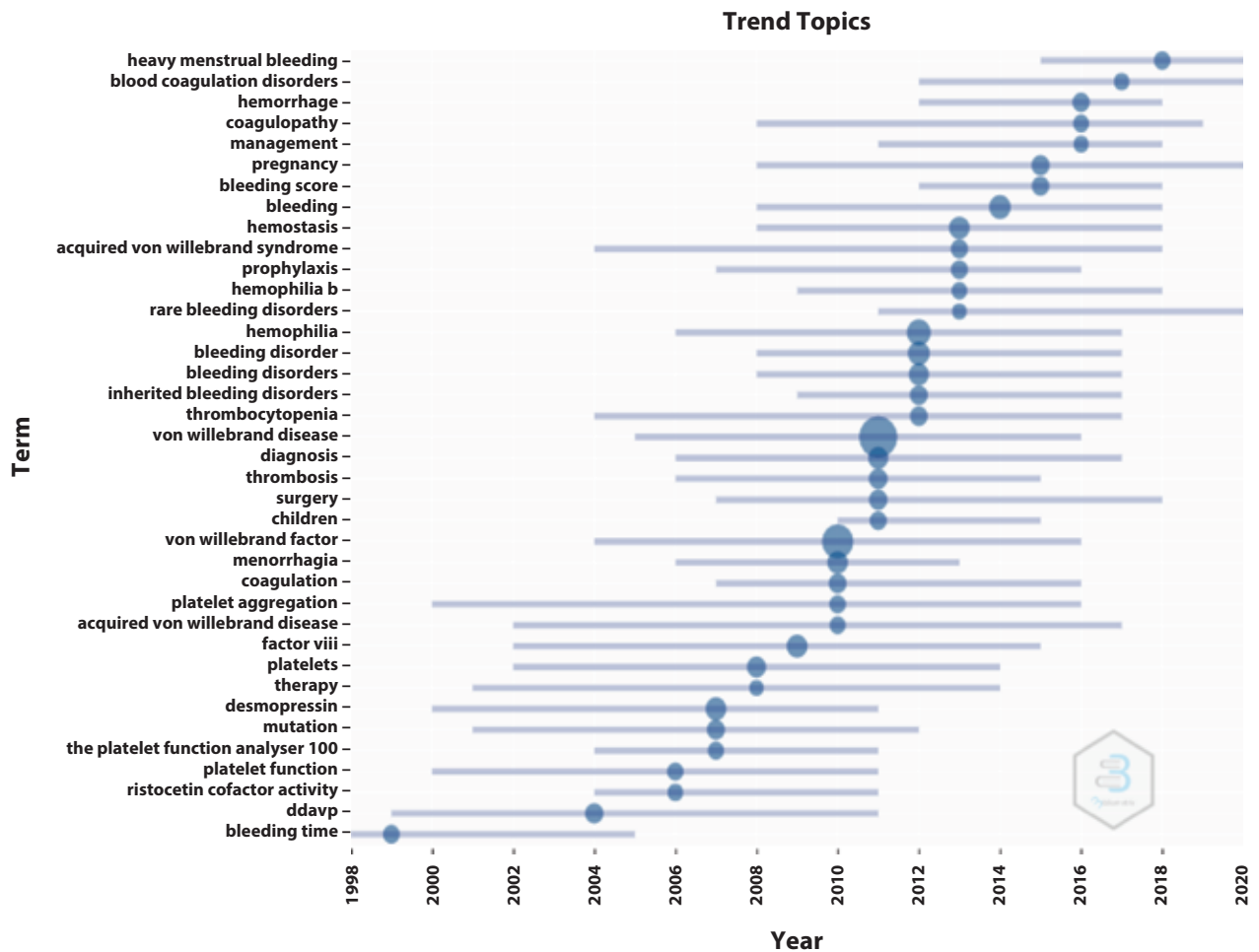


Figure 3. Trend topics.

recent publications has been observed (13-15). On the other hand, our data revealed that, although the number of publications increased from 2013 to 2021, the overall number of citations decreased within the same time span. Furthermore, the number of publications in 2021 was about double that of 2005; yet, the aggregate number of citations has declined dramatically throughout this time period. This implies that investigators interest on VWD has increased as the volume of papers published has increased; however, the quality of papers published may have decreased, as evidenced by a decreasing trend in citations and an increasing trend in non-cited papers from 2018 to 2021. Citations are determined by the quality of the work and the impact of the factors examined (16). The overall number of citations may also explain the influence among scholars

rather than the mainstream audience (17). Whereas the number of citations may reflect the impact in scholarly literature, citation count does not provide an indicator of an article's current relevance, which is best evaluated by the "total citations-per-year (CPY)" variable. According to our findings, Srivastava A had the greatest CPY, which was greater than the CPY of top publications in other domains such as valvular heart disease (median: 54) (15), sudden infant death syndrome (median: 12.1) (18), and hemorrhagic stroke (median: 17.5) (19). In our analysis, the most citations for a publication by Srivastava A was 1154, which is comparable to sepsis, where the highest number of citations for a paper was 1100 (20). Briefly stated, our findings indicate that there is a growing interest on VWD, with more scientists developing inquisitiveness



Figure 4. Most frequent keywords.

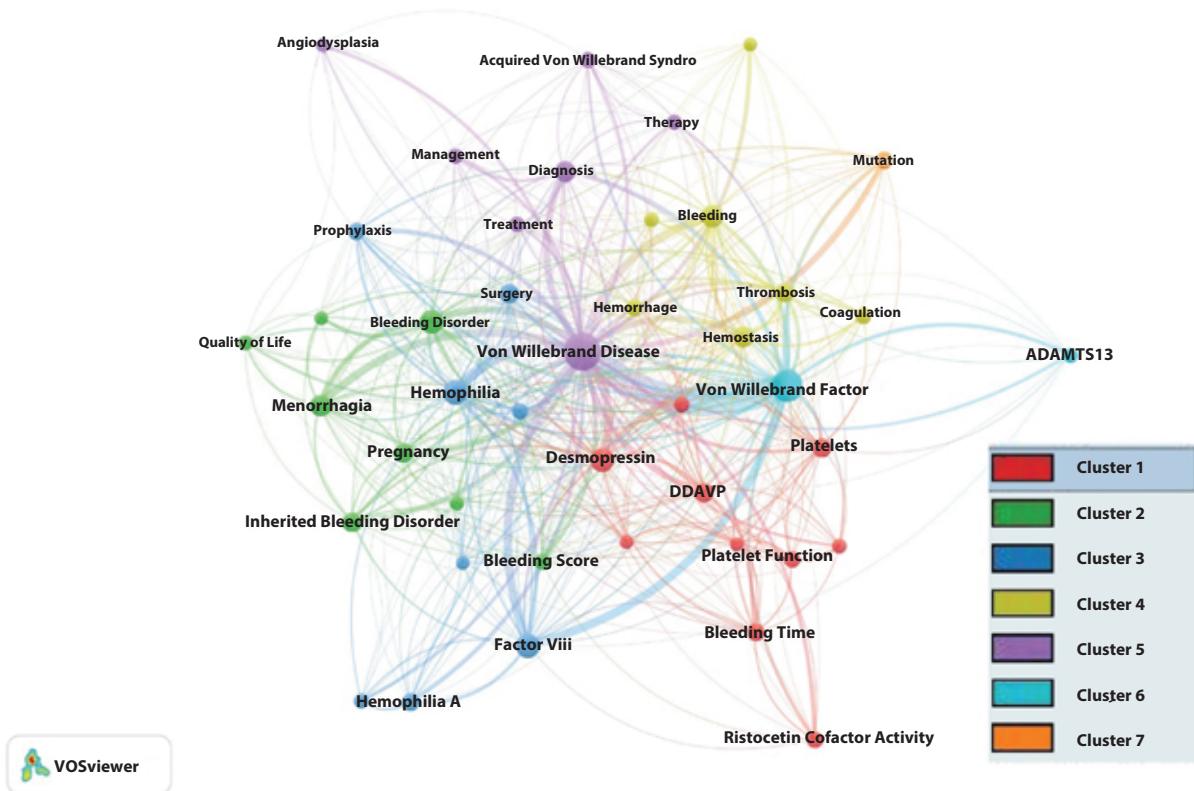


Figure 5. Author keywords.

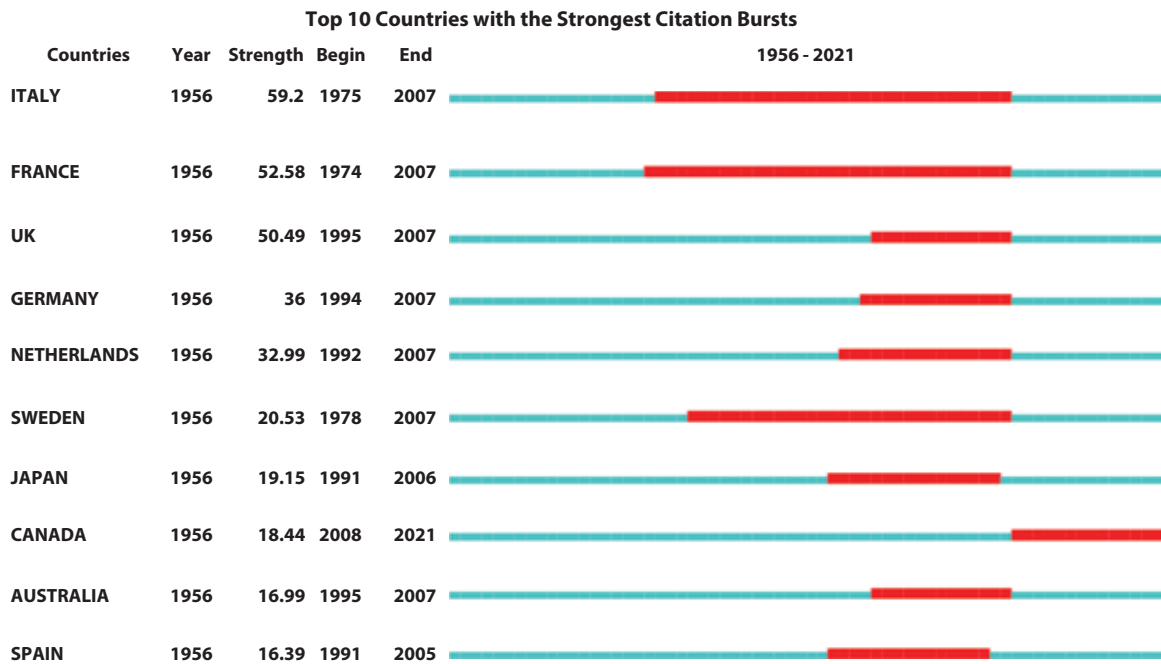


Figure 6. Countries with strongest citation bursts.

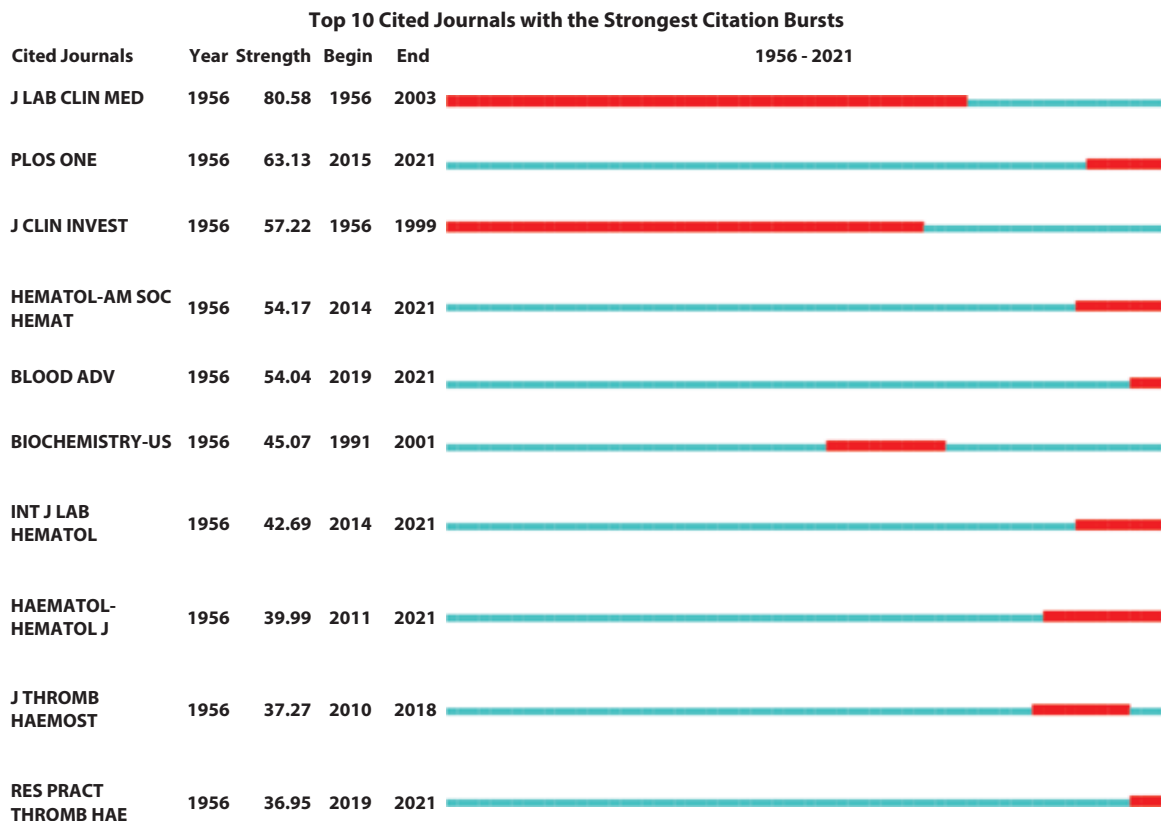


Figure 7. Cited journals with the strongest citation bursts.

for exploring further about the disease. It should be noted that while publications have increased significantly in recent years, obtaining citations takes considerable time and it may take several years to fully comprehend the influence of a recently published paper (17). The number of publications and citations received also depends on the indexing status of journals in different databases (21).

5.2 Most productive countries

The USA had the most publications (n=1048), followed by Italy, Germany, and the UK. Additionally, the USA had the highest number of citations (n=33512), followed by Italy, the UK, the Netherlands, and France. Similar findings have been observed in other bibliometric analyses (18,22,23). The healthcare system in the USA is well-funded, and regulations facilitate the production of research publications, which results in enormous contributions. Another metric that enables researchers to identify papers that have received a lot of attention in a short period of time is detecting citation bursts (24). The earliest citation was from France and it remained in rapid growth till 2007. Even though France had the longest period, Italy had the largest growth as compared to all the other countries in the group. Canada was the only country that started after 2008 and was at a high growth rate as of 2021. Remarkably, our findings indicate that the USA did not see any citation bursts.

The total publications and citations from Asian countries were 422 and 9919, respectively. In comparison to the USA, Asian countries had a very low total publication and total citation count. VWD is more prevalent in Asian countries like India and Taiwan than in Western countries like the USA (3). There may be a dearth of information on the incidence of VWD in certain countries resulting from misunderstandings and the absence of knowledge about inherited diseases (25). An Iraqi study on inherited bleeding disorders reported that more than 75% of patients had a family history of the consanguineous marriage of their parents and VWD was the most common type of inherited bleeding disorder observed (26). The lack of information may be attributed to a variety of elements, including the lack of diagnostic resources and

the lack of general awareness of the disease in the general public (27). When combined with the high incidence reported in Asian countries such as India, where relatively small studies have revealed a prevalence of 10 percent as opposed to 1 percent globally, it is apparent that immediate action is warranted to address this gap on a massive level (28,29). Future researchers may begin by examining the existing dearth of significant literature from Asian scholars, which represents a severe knowledge gap that must be closed. This can serve as a starting point for future research. Despite this fact, the top three Asian countries in terms of contributions were Japan, India, and Iraq.

5.3 Most productive organizations

In terms of contributing institutions, the 'University of Milan' produced the most research, with 192 publications and received 9841 citations, as highlighted in Table 7. Out of the top contributing organizations, 3 belonged to Italy. Italy has made progress and made attempts to improve diagnostic accuracy for patients with VWD by establishing registries and issuing new recommendations to address the shortage of diagnostic facilities for patients with VWD (30). Our analysis revealed that 'San Bortolo Hospital' with 92 publications received 6250 citations. The number of publications is not always proportional to the overall number of citations, as a high volume of publications is not always indicative of any particular institution's genuine influence. Thus, authors, institutions, and all other collaborators should not be merely concerned with the number of publications, but also with the impact and value that each publication offers.

5.4 Sources of publication

According to Bradford's law of scattering (31), the most influential authors obtain their citations from a small number of "core publications," and when they depart from these journals, their citations appear to diminish. These key journals are regularly cited in the literature of a certain discipline and hence provide significant value to scholars in that field. According to our research, the primary journals in the field of VWD are *Haemophilia*, *Journal of Thrombosis and Hemostasis*, and

Thrombosis and Haemostasis, as illustrated in Table 5. Although the journal, *Haemophilia*, has a lower impact factor than others, it appears to produce a significant number of citations for papers on this subject due to its specialized nature. Considering that studies on VWD are of particular interest to hematologists rather than the general medical audience, authors can ensure that their intended audience is reached by publishing in this journal. We discovered no significant link between a journal's impact factor and the number of most-cited articles published in that journal. For example, *Haemophilia* and the *Journal of Thrombosis and Haemostasis* have a lower impact factor than *Blood* yet have a comparable citation count. This indicates that high-quality articles on VWD are likely to have an influence independent of the journal's impact factor.

5.5 Most prolific authors

In our assessment, we identified that the top 10 authors published documents ranging from 65 to 119. The number of these documents published by the top 10 authors is higher when compared to another bibliometric analysis where the respective numbers ranged from 42 to 97 (32). We found 'Favaloro EJ' to be the most productive author, with 119 publications and 4394 citations, followed by 'Federici AB', as highlighted in Table 6. The larger number of prominent authors seen in our study suggests that a group of eminent experts is guiding the literature on VWD. A significant aspect of having an influential author on the team is that collaborating with an influential co-author can help young researchers advance their careers (33). Key author identification assists other scholars in the field in identifying prospective colleagues and mentors. In addition, it is critical to recognize and honor influential authors and their work in the age of evidence-based medicine. It has been demonstrated that scientists who make significant contributions to the literature are more likely to receive academic advancement, research grants, and favorable responses from journal editors (34). The number of writers has a significant impact on the overall citation count. By adding more authors, abilities and information expand, theoretically boosting the scientific quality of the paper. Moreover, having more contributors increases the visibility of a

document by exposing it to a wider network of professional contacts.

5.6 Top cited documents

The top 10 most cited papers were published between 1987 and 2013. The most cited paper was "Srivastava A, 2013, *Haemophilia*," which received 1154 citations and had a citation rate of 128.22 per year, as summarized in Table 4. However, it should be noted that, over time, publications may outnumber each other in terms of total citations. The top three most-cited publications were about the pathogenesis of VWD and management according to current guidelines. Although the citation count is not absolute, it is still the most widely used method of gauging academic impact by scholarly journals, individual academics, and funding organizations (16). Individual papers should receive citations based on the significance and utility of the results reported. Large enough data sets, on the other hand, show that there are criteria independent of individual article quality that can influence an average citation rate. With that stated, the citation count has been proposed as a major criterion for assessing the impact of research, which ultimately dictates the distribution of resources by funding organizations (35). However, it is still debatable whether citation frequency is important when allocating resources (36).

5.7 Network of keywords

Selecting the right keywords eases the identification of reports by other scholars (37). The most often listed term, as implied by the title, was 'Von Willebrand Disease,' as seen in Figure 4. However, since 'Von Willebrand Disease' is the term in the title as well, the next most often displayed term might be of greater significance. The subsequent most frequently listed keywords were von Willebrand factor, haemophilia, and bleeding disorder, indicating the pivotal role of publications linked with these keywords.

5.8 Limitations

In the current bibliometric analysis, a number of limitations should be noted, and the results should

be seen in the context of the methodology used. Firstly, many significant non-English articles may be missing due to the likelihood of English language bias in the database used. Secondly, our study can only offer hints about the trends of VWD research; it cannot provide conclusive proof for the complete body of literature on the subject. Thirdly, because we only used one database, bibliometric measurements (such as citation count) may differ from those found in other databases like Google Scholar and Scopus. To facilitate a fair comparison of articles, we opted to use a single database. Additionally, as in other bibliometric research, there is a possible length time-effect bias, which disadvantages more recent papers in terms of citations. Moreover, there is always the potential for self-citations, which leads to bias in bibliometric analysis, as these citations do not accurately reflect the paper's genuine "impact." Finally, restricting the search to the fields of title, abstract, and keywords may result in our search missing relevant publications.

6. Conclusions

The current study provided insights into the worldwide research and publication trends on VWD. The present analysis found that researchers are focusing attention on VWD research, as seen by the modest rise in publications throughout the years. Our study demonstrates that the USA had a considerably greater effect on VWD literature than Asian countries. Our analysis also reveals that the 'University of Milan' was the primary contributing organization, while 'Favaloro EJ' was the most significant author. *Haemophilia* was the leading journal on the subject of VWD. The data reported here may be valuable in guiding researchers as they plan future paths in the hope that the field of VWD becomes more productive. Efforts should be made to boost funding and support research in areas with a scarcity of published material.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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