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Dandy–Walker syndrome: a bibliometric analysis of the most 100 cited articles

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Introduction: Dandy–Walker syndrome (DWS), a complex neurodevelopmental disorder, has intrigued clinicians and researchers since its description by physicians Walter Dandy and Arthur Walker. Despite its recognition for nearly a century, understanding its etiology, pathogenesis, and clinical manifestations remains elusive. This bibliometric analysis aims to elucidate influential academic works on DWS.

Methods: In January 2024, the authors conducted a Scopus search for articles on DWS and identified the top 100 referenced publications. The Harzing Publish or Perish search engine was utilized with relevant terms, including 'Dandy-Walker', 'Dandy-Walker Syndrome', and 'Dandy-Walker Malformation'. Data from Scopus, including publication details and citation counts, were compiled and organized using Microsoft Excel. Statistical analysis and data visualization were performed using Python, with Pandas, Matplotlib, Seaborn, and NetworkX libraries employed for this purpose.

Results: The bibliometric analysis of DWS research revealed key insights. Significant research output was noted in the 2000–2009 and 1990–1999 decades. The cumulative citations totaled 6059, with an average of 2.60 citations per year per article. Leading authors included W B Dobyns, Kathleen J Millen, and G Pilu. Institutions such as the University of California and Harvard Medical School were prominent, with the United States being the predominant contributor. Major journals like the American Journal of Medical Genetics played significant roles.

Conclusion: This bibliometric study summarizes the most-cited articles on DWS, providing light on the field and its seminal works that have shaped both present-day clinical treatment and the trajectory of future research.

Keywords: Dandy–Walker-Syndrome, Dandy–Walker-Malformation, fourth ventricle

Introduction

Dandy–Walker syndrome (DWS) is a complex and sophisticated neurodevelopmental condition that attracts the interest of physicians, academics, and scientists. The disease, which was initially documented by physicians Walter Dandy and Arthur Walker in the early 1900s, encompasses a range of congenital abnormalities that impact the cerebellum and its surrounding structures[1]. The medical profession still faces hurdles in understanding the causes, development, and clinical symptoms of DWS, despite its awareness for almost a century.

Cystic dilatation of the fourth ventricle, agenesis or hypoplasia of the cerebellar vermis, and an expanded posterior fossa are the defining signs of DWS[2]. These structural abnormalities frequently result in a wide range of neurological and developmental problems, including motor deficiencies and cognitive and psychosocial challenges[3]. Understanding the underlying mechanisms and exploring new therapy methods is crucial due to the varied clinical manifestations of this condition, which adds to its enigmatic nature.

HIGHLIGHTS

- This bibliometric analysis aims to elucidate influential academic works on DWS.
- In January 2024, we conducted a Scopus search for articles on DWS and identified the top 100 referenced publications and found that he cumulative citations totaled 6059, with an average of 2.60 citations per year per article.
- In our article, we summarize the most-cited articles on DWS, providing light on the field and its seminal works that have shaped both present-day clinical treatment and the trajectory of future research.

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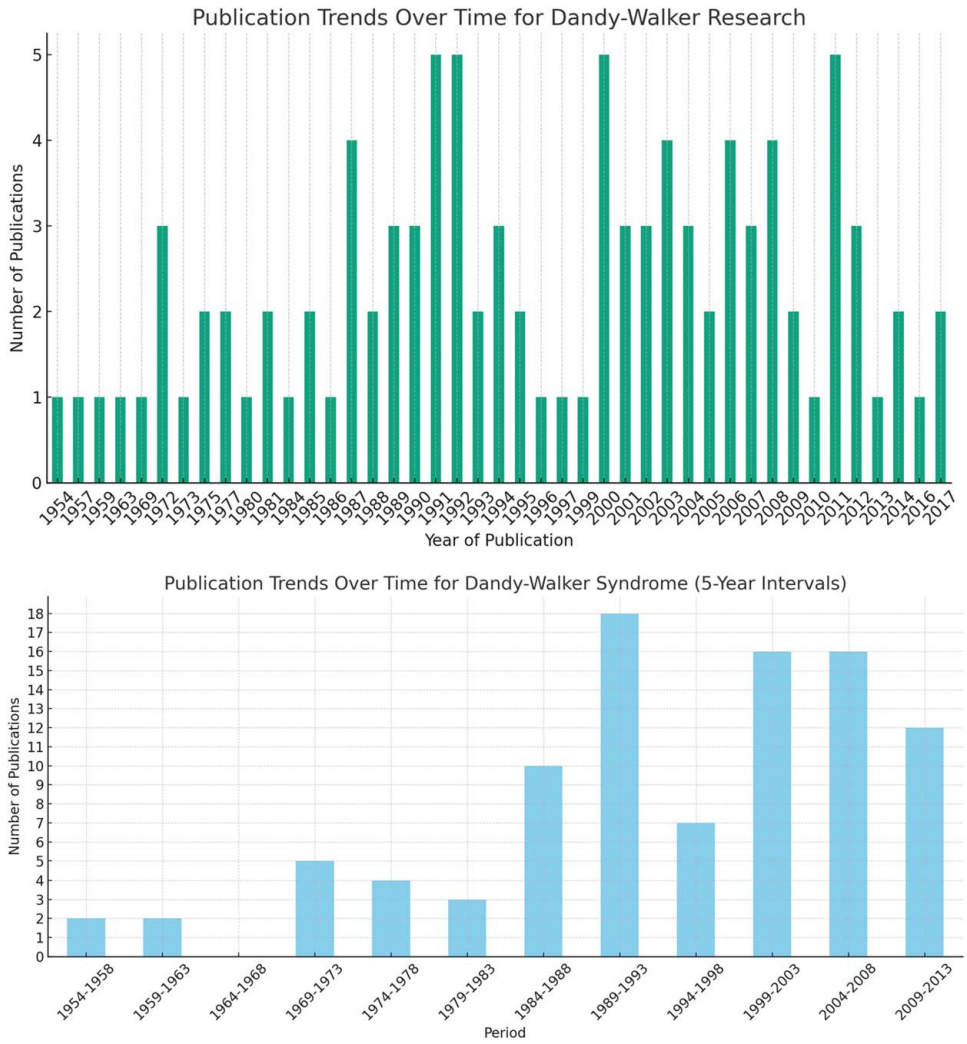

Figure 1. Trends in scholarly publications on Dandy–Walker syndrome. Top: yearly from 1954 t0 2017. Bottom: grouped into 5-year intervals from 1954 to 2013.

We aim to provide a bibliometric analysis of the most influential academic works concerning the DWS and to highlight the main topics that had an impact on current clinical practice.

Methods

An evaluation of the Scopus collection was conducted in January 2024 to ascertain the number of citations received by all published publications pertaining to DWS. We ascertain the 100 most often cited publications on this topic. The Harzing Publish or Perish search engine was employed to query the keywords 'Dandy-Walker', 'Dandy-Walker Syndrome', and 'Dandy-Walker Malformation'.

The inclusion criteria encompassed peer-reviewed papers specifically focused on Dandy–Walker Malformation, including original research articles, review articles, and case reports. The exclusion criteria encompassed articles pertaining to patents, books, chapters, manuals, and nonbiomedical publications.

The initial screening process entailed evaluating the suitability of study titles and abstracts. Two separate authors (O.A. and Y. A.) conducted a comprehensive review to find the publications with the highest number of citations, after eliminating duplicates. Later on, the entire article was obtained. If there were any inconsistencies, a third reviewer (B.C.) was consulted to make the final decision. The data underwent comprehensive analysis, including several variables like publication titles, publication years, citation counts, authorship, institutional affiliations, geographical origins, journal names, study categories, and demographic characteristics.

We employed Microsoft Excel (version 16.81) for initial data organization and cleaning. For the statistical analysis and data visualization, we utilized Python programming language, specifically leveraging libraries such as Pandas for data manipulation, Matplotlib, and Seaborn for creating informative visualizations, and NetworkX for network analysis of author collaborations.

Results

Publication trends over time

The bibliometric analysis of DWS research revealed significant

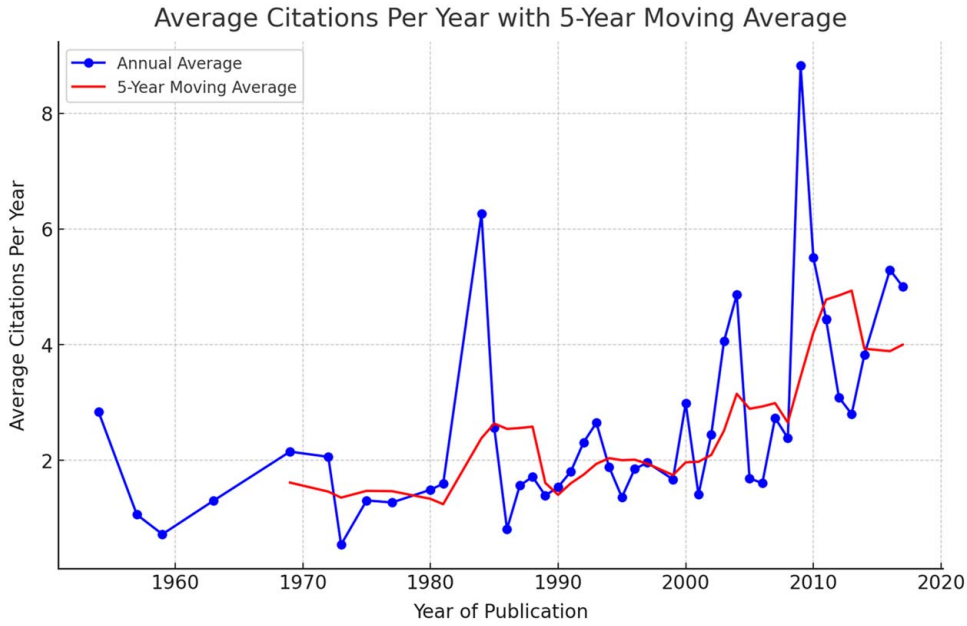

Figure 2. Evolution of average citations per year in Dandy–Walker syndrome research publications. The blue line represents the annual average citations per article, while the red line shows the 5-year moving average, illustrating a smoothed overview of research interest and impact over time.

insights into the research trends over the years. Figure 1 provides a clear picture of the evolving academic interest in this field, depicting the number of publications per year. The data shows fluctuations in research output, indicating periods of heightened focus and research activity. Notably, the decades from 2000 to 2009 and 1990 to 1999 saw the most significant research output, with 33 and 21 highly cited articles published, respectively.

Citations

Our citation analysis of the DWS research publications yielded insightful findings regarding their impact over time. The cumulative number of citations across all articles in our dataset totaled 6059, reflecting the broad influence of these works in the field. On average, each article garnered about 2.60 citations per year, indicating a steady recognition of these publications in the academic community. The line graph presented in Figure 2 illustrates the trend of average citations per article over time, enhanced by a 5-year moving average. This representation smooths out short-term fluctuations, providing a clear view of the sustained academic interest in DWS research through the decades. Additionally, our analysis identified the top 10 articles based on citations per year, offering a more equitable measure of impact over time. The 2009 publication, 'FOXC1 is required for normal cerebellar development and is a major contributor to chromosome 6p25.3 Dandy–Walker malformation,' led with an average of 15.36 citations per year, highlighting its significant influence since publication. This was followed by the 2004 study, 'Heterozygous deletion of the linked genes ZIC1 and ZIC4 is involved in Dandy–Walker malformation,' which accumulated an average of 9.79 citations per year, as shown in Table 1 and Figure 3.

Authorship analysis

The authorship analysis of the DWS research publications yielded significant findings regarding the key contributors and collaboration patterns in this field. We identified the most prolific authors based on the frequency of their publications. Leading this list is W B Dobyns, with a total of six publications, followed by Kathleen J. Millen with five publications, and G Pilu with four publications. Other notable contributors include B.S. Mahony, Kimberly A. Aldinger, and D.G.

Table 1

The top five most cited-per-year articles on Dandy–Walker syndrome research.

Position	Title	Year of publication	Total number of citations	Citation per year
1	FOXC1 is required for normal cerebellar development and is a major contributor to chromosome 6p25.3 Dandy–Walker malformation	2009	215	15.36
2	Heterozygous deletion of the linked genes ZIC1 and ZIC4 is involved in Dandy–Walker malformation	2004	186	9.79
3	Dandy–Walker malformation: prenatal diagnosis and prognosis	2003	164	8.2
4	The Dandy–Walker malformation. A review of 40 operative cases	1984	243	6.26
5	Multiple developmental programs are altered by loss of Zic1 and Zic4 to cause Dandy–Walker malformation cerebellar pathogenesis	2011	71	5.92

Figure 3. Evolution of average citations per year in Dandy–Walker syndrome research publications. Top chart shows ranking based in cumulative number of citations; bottom chart shows the ranking based on citations per year.

Table 2

The most prolific authors based on the frequency of their publications on Dandy–Walker syndrome.

#	Author	Number of publications
1	W.B. Dobyns	6
2	Kathleen J. Millen	5
3	G. Pilu	4
4	B.S. Mahony	3
5	Kimberly A. Aldinger	3
6	D.G. Mclone	3
7	L. Bovicelli	2
8	J.R. Siebert	2
9	D. Parisot	2
10	A. Mohanty	2
11	B.L. Maria	2
12	I.J. Frieden	2
13	W.J. Gardner	2
14	D.H. Ledbetter	2
15	F. Brunelle	2
16	R.P. Kapur	2
17	B.R. Benacerraf	2
18	J.A. Estroff	2
19	O. Klein	2
20	Inessa Grinberg	2

Mclone, each with three publications, as detailed in Table 2. Furthermore, the co-authorship analysis, visualized in Figure 4, depicts the interconnectedness and collaborative efforts among authors, revealing a highly interconnected community of authors, with W.B. Dobyns emerging as a pivotal figure.

Institutional and geographic analysis

Our analysis also revealed significant insights into the institutional and geographic distribution of DWS research. Figure 5 shows that among research institutions, the University of California and Harvard Medical School are leading the field, each contributing five publications. They are closely followed by Children's Hospital Boston, Hôpital Necker Enfants-Malades, Northwestern University Medical School, and The University of Chicago. Geographically, the United States stands out as the predominant contributor, with 43 publications, followed by France with 8, Italy with 6, Turkey with 5, and Greece with 3 publications.

Journal analysis

The study also focused on identifying the journals most frequently publishing research on DWS. The American Journal of Medical Genetics leads with 10 publications, indicating its significant role in disseminating research related to Dandy–Walker

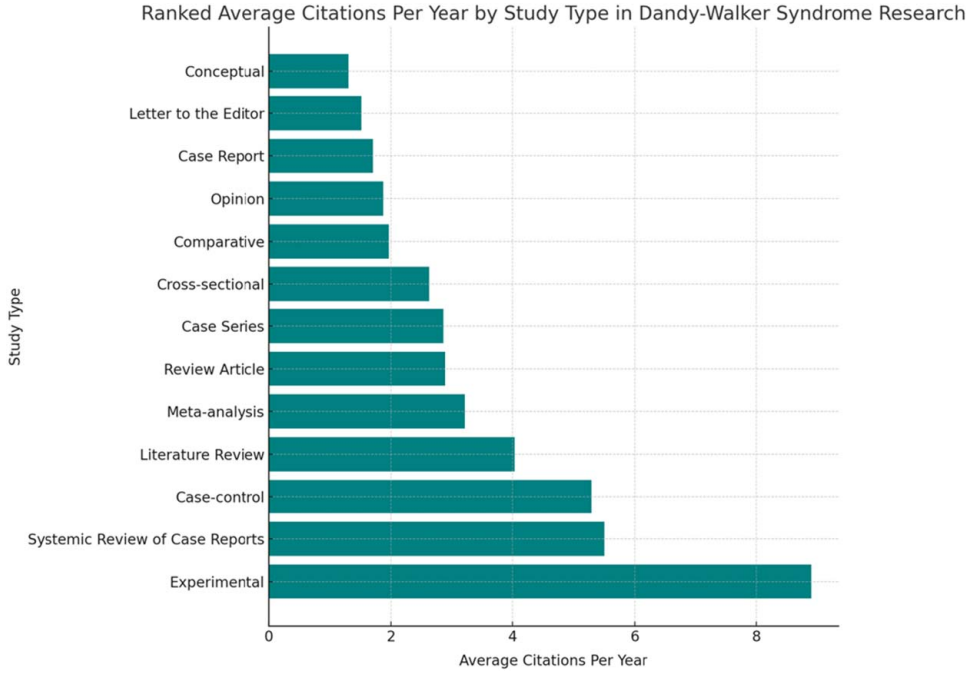

Figure 4. Average Citations Per Year by Research Methodology. Co-authorship network in Dandy–Walker syndrome research. Node color and size.

syndrome. Child's Nervous System and Ultrasound in Obstetrics and Gynecology follow with five and four publications, respectively. prenatal diagnosis and pediatric neurosurgery each contributed three publications in this field.

Study type distribution

Our analysis of the distribution of study types in DWS research reveals the prevalent research methodologies. Figure 6 illustrates the frequency of various types of studies, with case reports and retrospective case series are the most common types of studies, with 46 and 38 publications, respectively. Other study types such as review articles, experimental studies, and meta-analysis studies are less frequent, with six, four, and one publication, respectively.

Population characteristics

Our analysis of the study population characteristics provides insight into the demographics most frequently studied. Figure 7 delineates the frequency with which various population groups are the focus of research in this field. Predominantly, research has concentrated on human children, with 79 studies targeting this demographic. This is followed by studies involving human adults and human fetuses, with 20 and 13 publications, respectively. Research on animals is less prevalent, with only five publications.

Discussion

Publication trends over time

The bibliometric analysis of the most cited literature on DWS offers a unique perspective into the evolution of research interests and the impact of scientific contributions in this field. The publication trends observed over time, as illustrated in Figure 1, not only reflect the fluctuating academic attention to DWS but also signal key periods of intensified research activity. Particularly, the increased number of publications in the decades from 2000 to 2009[4–36] and 1990 to 1999[37–57] indicates a growing research interest and potentially aligns with significant advancements in understanding or treatment modalities of the syndrome. This surge in research output could be attributed to evolving diagnostic capabilities, such as the advent of more sophisticated imaging techniques, heightened awareness of the syndrome's clinical implications, or increased funding and interest in this topic.

Citation

The citation analysis, encapsulating a total of 6059 citations across the dataset, highlights the broad influence and enduring relevance of the works in the field of DWS. The average citation rate per year suggests a steady acknowledgment of these publications within the academic community. The citation trends, as depicted in Figure 2, offer insight into the changing dynamics of research impact over time. The identification of the cumulatively most cited articles, with a leading article from 1984 garnering the highest number of citations, highlights seminal works that have significantly shaped current understanding and management approaches of DWS[58]. This pivotal study of 1984 by Hirsch.[58] meticulously analyzed 40 cases of DWS over 13 years. It notably identified a 2.4% incidence of DWS in hydrocephalus patients and highlighted the prevalence of postnatal hydrocephalus as a complication rather than an inherent part of DWS[58]. The study emphasized the criticality of distinguishing DWS from similar conditions and explored its pathogenesis, suggesting its onset between the 3rd and 4th postovulatory weeks. Importantly, the research delved into the associated brain and systemic malformations, providing a holistic view of the condition[58]. In terms of treatment and prognosis, it evaluated the effectiveness of cyst-peritoneal

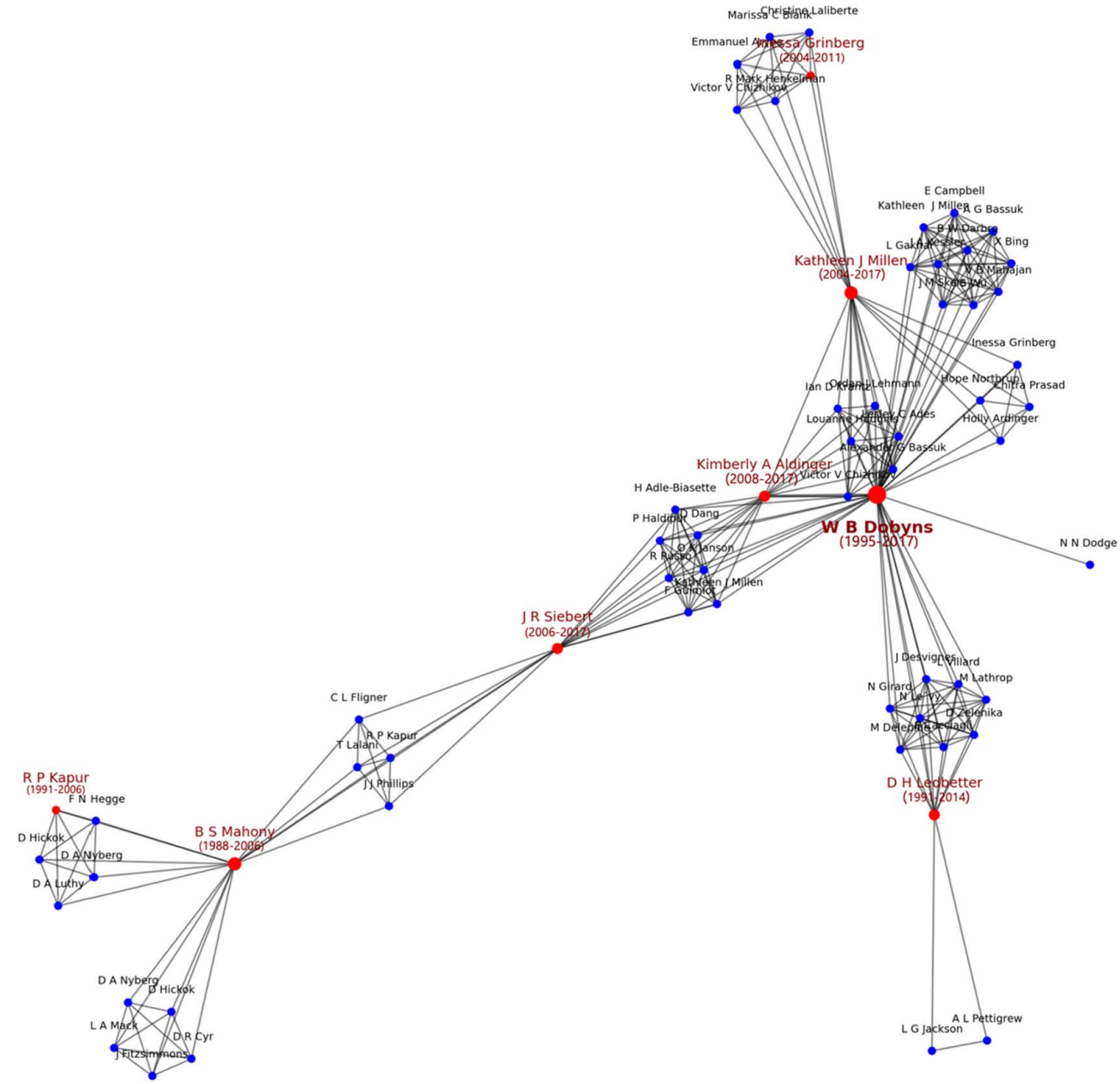

Figure 5. Top five institutions and countries by number of publications. Co-authorship Network in Dandy-Walker Syndrome Research. Node color and size: Red Nodes represent some of the top 20 authors in the Dandy-Walker Syndrome research field, Blue Nodes indicate all other authors included in the study. These authors, while not among the top 20, play important roles in the research network through their contributions and collaborations. The size of each node corresponds to the author's degree of connectivity within the network. Larger nodes indicate authors with a higher number of publications or greater involvement in collaborative research efforts. Node Annotations: Each red node is annotated with the range of publication years, denoting the active period of each significant author in the network. This provides a temporal dimension to the network, helping to visualize the span of research activity and collaboration over time. Edges: The lines connecting the nodes represent co-authorship links. Each edge signifies that the connected authors have collaborated on at least one research paper. Network Layout: The arrangement of the nodes is based on a force-directed layout, where authors who have co-authored, papers are positioned closer together, forming clusters. These clusters potentially represent research groups or collaborative communities within the field.

shunting and reported a 12.5% mortality rate, with 60% of patients exhibiting an intelligence quotient above 80[58]. This article has been instrumental in shaping the current understanding and management approaches of DWS, offering valuable insights into its clinical presentation, differential diagnosis, and therapeutic strategies.

This observation, from finding the most cited articles, highlights the enduring influence of foundational research while also acknowledging the significant impact of more recent studies, such as the 2009 publication by Aldinger *et al.*[35], noted for its high citation rate. This study presents significant findings in the understanding of DWM[35]. The study emphasizes the role of the

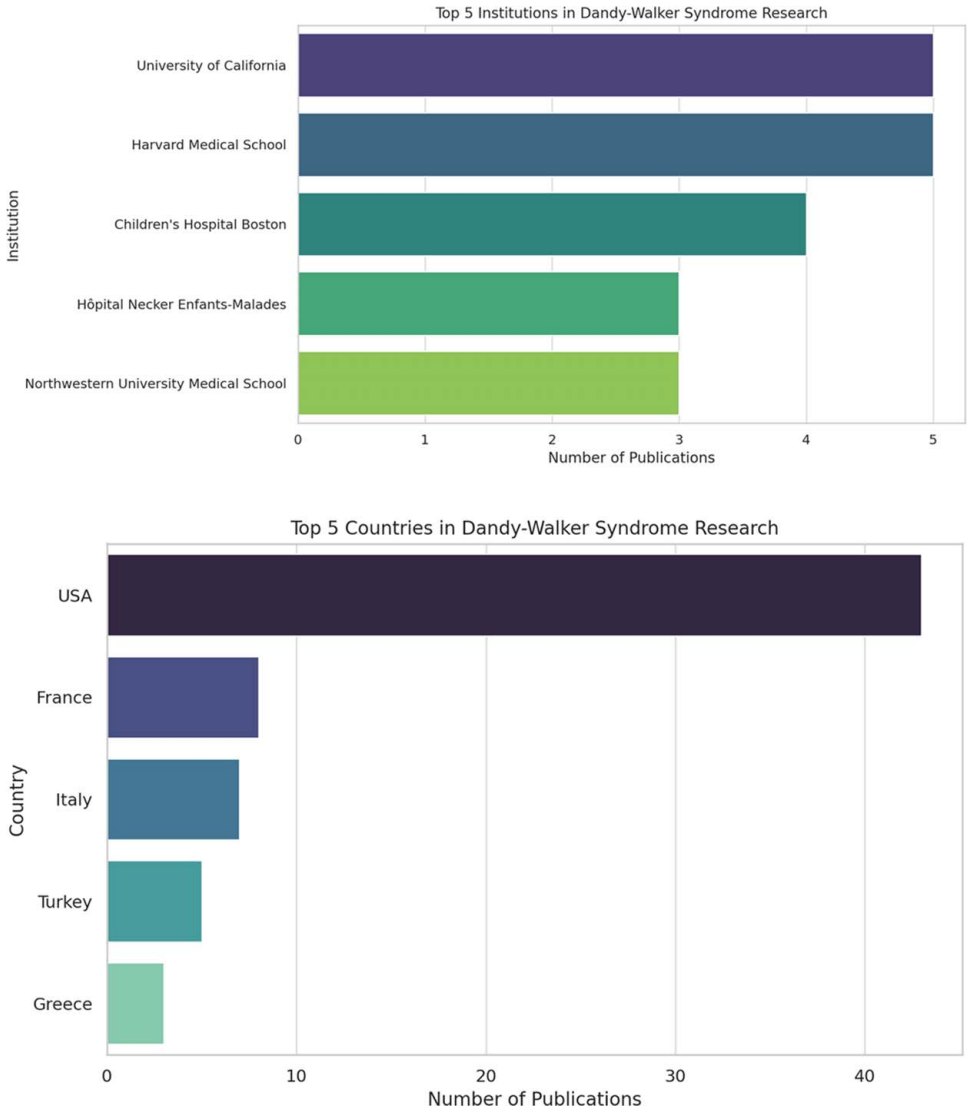

Figure 6. Frequency distribution of different study types in Dandy–Walker syndrome research.

FOXC1 gene in cerebellar development and its association with DWM[35]. It explores genetic variations like deletions or duplications in chromosome 6p25.3 and their impact on the cerebellum and posterior fossa malformations[35]. The research establishes a connection between human FOXC1 mutations and cerebellar vermis hypoplasia (CVH), contributing to a deeper understanding of DWS's pathogenesis and the interplay of genetic factors in its manifestation[35]. This study is crucial in the field for linking specific genetic alterations to DWS, paving the way for more targeted research and potentially improved diagnostic and therapeutic approaches.

Furthermore, the article titled Heterozygous deletion of the linked genes ZIC1 and ZIC4 is involved in DWS by Grinberg *et al.*[20] 2004, presents groundbreaking findings on the genetic aspects of DWS. The study focuses on the role of the ZIC1 and ZIC4 genes, located on chromosome 3q2, in the development of DWS[20]. Through detailed analysis, the research demonstrates that heterozygous deletion of these genes leads to DWS, backed by evidence from physical mapping of interstitial deletions in individuals with DWS and mouse model studies. The study also investigates the cerebellar vermis hypoplasia and the upward rotation of the cerebellar vermis in individuals with DWS[20]. This research was pivotal in identifying the first critical region associated with DWS and provides a foundation for understanding the genetic basis of DWS[20]. It emphasizes the importance of these genes in cerebellar development and their potential as targets for future therapeutic strategies. The findings have significant implications for the diagnosis, management, and further research into DWS, highlighting the complexity of genetic factors involved in this condition.

A critical observation emerged from calculating the data to reflect the average number of citations per year for each research methodology, ensuring a balanced assessment across publications of varying ages. As depicted in Figure 8 and detailed in Table 3, this approach reveals a landscape of research impact. The analysis shows that certain study types, particularly experimental studies, which lead with the highest average citations per year, and systemic reviews of case reports, are more frequently

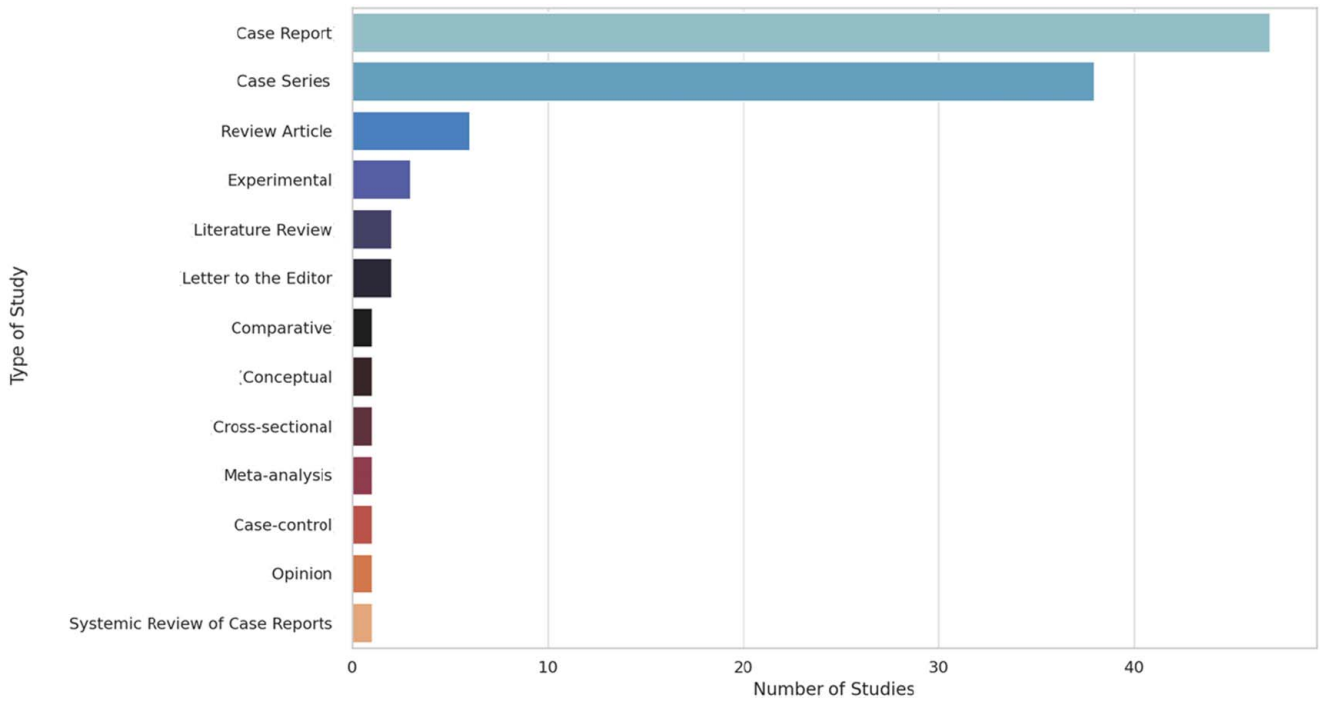

Figure 7. Study population distribution in Dandy–Walker syndrome research.

cited on an annual basis. Notably, the 2009 study of Aldinger *et al*. and the 2004 study by Grinberg *et al*. discussed above for being in the top five most cites publications, were experimental studies[20,35]. The trend of experimental studies receiving higher citations suggests that these methodologies, despite being less frequent in the corpus, resonate strongly within the academic community, potentially due to their novel insights to the subject matter.

On the other hand, more frequently observed study types like case reports and retrospective case series reviews, despite their utility in clinical and observational insights, exhibit a comparatively lower average citation count. This disparity highlights the diverse nature of research contributions and their varying receptions in the scientific community. Such insights are invaluable for researchers, indicating potential areas for future research focus and publication strategies. Our analysis highlights the multifaceted nature of research impact, emphasizing that frequency of publication does not always correlate with citation impact, thereby enriching our understanding of the research dynamics in this field.

Authorship analysis

In examining authorship patterns, our findings reveal key contributors to the DWS research field. The prominent role of authors illustrated through their extensive publication record and central position in the

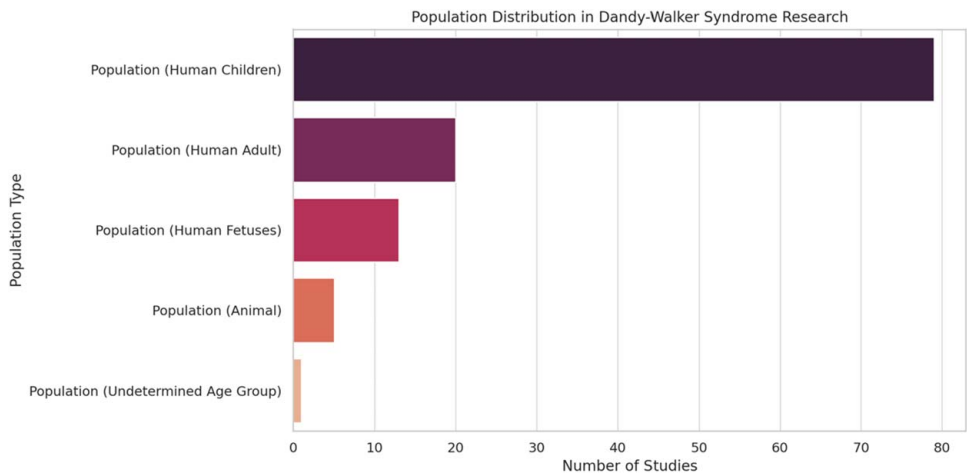

Figure 8. Average citations per year by research methodology.

Table 3

Average citations per year by research methodology in Dandy–Walker syndrome research.

Rank	Study type	Average citations per year
1	Experimental	8.89
2	Systemic review of case reports	5.50
3	Case–control	5.29
4	Literature review	4.04
5	Meta-analysis	3.22
6	Review article	2.89
7	Case series	2.85
8	Cross-sectional	2.62
9	Comparative	1.96
10	Opinion	1.87
11	Case report	1.70
12	Letter to the Editor	1.52
13	Conceptual	1.30

co-authorship network (Fig. 4), highlights individuals who have significantly influenced the trajectory of research in this area.

This visualization reveals a highly interconnected community of authors, with W.B. Dobyns emerging as a pivotal figure. The prominent positioning and size of the node representing W.B. Dobyns in the network are indicative of their significant influence. This prominence is attributable not only to their high publication count but also to their role as a crucial collaborative link connecting various research clusters[20,35,55,59–61]. The network clearly delineates W.B. Dobyns as not just a prolific contributor but also as a central node facilitating extensive collaborative ties across different research groups. This finding highlights the integral role played by W.B. Dobyns in advancing the field of DWS research.

Within the network, distinct clusters emerge, likely representing specific research groups or collaborative communities. The top 20 authors, some of which are distinguished in red, are integral to these clusters (Fig. 5). They demonstrate multiple interconnections within and across these clusters, indicating a robust collaborative network. For instance, authors like Kathleen J. Millen, exhibits significant collaborative roles, as evidenced by their numerous connections within the network. W.B. Dobyns and Kimberly Aldinger are co-authors with Kathleen J. Millen of the one of the most cited publications, the FOXC1 study discussed previously[35]. This shows that the role of those authors is not only significant in terms of research networking, but also highlights their works that have significantly improved our understanding of DWS. Other notable contributors, such as B.S. Mahony, serves as key links between different authors and groups, further emphasizing the collaborative nature of scientific research in this domain[27,41,62].

The network's layout also reveals important cross-group collaborations, where authors like W.B. Dobyns act as bridges between disparate research clusters. These intercluster connections are crucial in fostering a holistic and integrated approach to research, promoting the exchange of diverse ideas and methodologies. This aspect of the network highlights the collective effort and interconnectedness inherent in scientific research, particularly in complex fields.

This co-authorship network visualization transcends mere data representation, unfolding a narrative of collaboration and intellectual synergy. It serves as a roadmap for new researchers to identify key contributors and potential collaborators. For academic institutions and funding bodies, it highlights the critical role of collaborative networks in driving scientific innovation and progress. The network not only reflects the current state of research in the field but also offers insights into its evolution, showcasing how new contributors integrate into existing networks or establish new collaborative entities.

Institutional and geographic analysis

The institutional and geographic analysis points to the significant contributions from renowned research centers and highlights the global scope of DWS research. The leading role of institutions like the University of California and Harvard Medical School, as well as the predominant contribution from the United States, indicates concentrated centers of research excellence. However, the presence of contributions from countries like France, Italy, Turkey, and Greece highlights the worldwide interest and collaborative efforts in understanding and addressing DWS[4,5,8,15,16,18,21–23,29,36,39,49,58,59,63–67]. This geographic diversity is crucial for a comprehensive understanding of the syndrome, considering potential regional variations in its presentation and management.

Journal analysis

Furthermore, the journal analysis reveals that specific academic journals have played a pivotal role in disseminating research on DWS. The prominence of journals such as the American Journal of Medical Genetics in publishing studies related to this syndrome suggests their critical role as platforms for advancing understanding and treatment approaches. The diversity of journals, ranging from genetics to neurosurgery, highlights the multidisciplinary nature of DWS research. This diversity is vital for fostering a holistic understanding of the syndrome, encouraging cross-disciplinary collaboration and integration of knowledge from various medical and scientific domains.

Following the American Journal of Medical Genetics, Child's Nervous System has contributed five publications, and Ultrasound in Obstetrics and Gynecology has published four articles. Prenatal diagnosis and pediatric neurosurgery each have three publications in this field. The frequency of publications in these journals not only highlights their focus on DWS but also their influence in the broader research community studying this condition. The prominence of these journals in this specific area suggests their importance as platforms for the latest research and developments in the field of DWS. This is beneficial for researchers looking for the most relevant and impactful journals for their work in this area.

Study type distribution

The study type distribution analysis offers an insightful perspective into the methodologies prevalent in DWS research. The dominance of case reports and retrospective case series reviews, as shown in Figure 6, indicates a research trend focused on clinical observations and historical data analyses. While these study types are invaluable for gathering detailed clinical information and forming hypotheses, the relative scarcity of experimental, longitudinal, and comparative studies points to a potential gap in the field. The relatively lower frequency of experimental studies may point to an opportunity for future research efforts to focus more on experimental approaches, which could provide new insights into the etiology and treatment of this condition. This

study offers a comprehensive view of the research methodologies employed in DWS studies, helping to identify prevailing trends and potential gaps in research methodologies.

Population characteristics

The analysis of study population characteristics, as illustrated in Figure 6, emphasizes the focus on pediatric populations in DWS research. This aligns with the congenital nature of the syndrome and highlights the importance of early diagnosis and intervention. However, the lesser emphasis on adult populations and fetuses in the research highlights an area for further exploration. Understanding the long-term outcomes and the prenatal aspects of DWS is essential for comprehensive patient care and management. Moreover, the inclusion of animal studies, although limited, points to the potential for experimental and pathophysiological research that can complement clinical studies and contribute to a more profound understanding of the syndrome[20,35].

Limitations

Bibliometric analyses, while powerful in synthesizing and evaluating academic literature, inherently possess certain limitations, some of which were encountered in our study on DWS[68,69]. A primary limitation lies in the dependency on the accuracy and completeness of the databases used[70]. Misclassifications, incomplete data, or omissions in these databases could lead to skewed results. Our study relied on datasets from reputable academic databases, yet there is always a risk of inherent biases in these sources, including the potential omission of influential works that are not indexed.

Furthermore, citation-based analyses can sometimes favor older publications simply because they have had more time to accumulate citations[71,72]. While we attempted to mitigate this by considering average citations per year, the inherent advantage of time for older articles remains a factor. Additionally, citation counts do not always equate to research quality or impact, as citations can be influenced by various factors, including journal visibility, networking among researchers, and trends in research topics.

Conclusion

Our comprehensive bibliometric analysis of DWS research provides a panoramic view of the landscape of scholarly work in this field. We have identified key trends in publication, influential contributors, and pivotal institutions driving the research. The fluctuations in research output over time and the identified citation trends offer valuable insights into the evolving academic interest and impact of studies in this field. The prominence of certain journals and the diversity of study types highlight the multidisciplinary nature of research on DWS. Our findings not only reflect the historical and current state of research but also illuminate potential areas for future investigation, particularly in underrepresented methodologies and demographics. Our study highlights the importance of continuous exploration, interdisciplinary collaboration, and the integration of diverse research methodologies to advance our understanding and management of this complex condition. As we move forward, it is essential to build upon these foundations, embracing both the breadth and depth of research to pave the way for innovative approaches and improved patient outcomes in DWS.

Ethical approval

Ethics approval was not required for this review.

Consent

Informed consent was not required for this review article.

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Author contribution

All authors have contributed equally in formation of all forms of manuscript.

Conflicts of interest disclosure

The authors declare that they have no financial conflicts of interest with regard to the content of this report.

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Unique identifying number or registration ID: not applicable.
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Guarantor

Bipin Chaurasia.

Data availability statement

Not applicable.

Provenance and peer review

Ok.

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