The Top-100 most cited articles on Moyamoya disease: A bibliometric analysis

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Objective: Moyamoya disease (MMD) is a progressive steno-occlusive cerebrovascular phenomenon with unknown pathogenesis. Considering the abundance of articles addressing Moyamoya disease, a detailed analysis concerning the publication trends is of paramount importance. The aim of the study is to report the current knowledge of the top-100 most cited articles on Moyamoya disease in the literature.

Methods: A non-time restricted keyword-based search was performed in June 2020 using the Scopus database. The search keywords included the following: “Moyamoya”, “Moyamoya disease”, and “Moyamoya syndrome”. The search result was used to rank the articles based on their citation count. The top-100 most-cited articles were obtained and classified into seven categories.

Results: A total of 3,543 articles on Moyamoya disease were published between 1955 and 2020. The Top-100 articles were published between 1977 and 2016 with a total of 16,119 citations, per year, and 7.23% rate of self-citation. The 1990s was the most productive decade (N=42). The most contributing country to the list was Japan (N=60). Stroke was the most active journal (N=23). Houkin, K., a Japanese neurosurgeon, was the most prolific author (N=15).

Conclusions: Moyamoya disease has been extensively investigated in the literature throughout the years. The majority of articles published in the literature were addressing the surgical management and clinical outcome. Authors from neurosurgical backgrounds were the most active contributors to the field of Moyamoya disease.

Keywords Moyamoya disease, Moyamoya syndrome, Stroke
INTRODUCTION

Moyamoya disease (MMD) is an uncommon cerebrovascular abnormality with a pathognomonic feature of progressive occlusion of supraclinoid internal carotid artery (ICA). Takeuchi and Shimizu were the first to describe this abnormality in 1957 as bilateral hypoplasia of the ICAs where Kudo T. was the first to describe it as a spontaneous occlusion of the circle of Willis in 1968.24(41) Moyamoya, a Japanese term, was first introduced to the medical literature by Suzuki and his colleague Takaku in 1969.40 The term characterizes the obscured hazy angiographic appearance of collaterals “MoyaMoya vessels (; puff of smoke)” resulting from insufficiency in the circle of Willis.35(40) The epidemiological patterns of Moyamoya reveal its predominance in Eastern Asia.20 Bimodal peaks of age have been noted with the disease distribution, particularly in the first and fourth decades, and it is slightly more common in females.20 Although more than half a century has passed since the first time a Moyamoya case was reported in 1957, the etiology of Moyamoya is not yet well established, and many incongruities in its pathogenesis are still unresolved.41 The diagnostic criteria for MMD according to Fukui et al. involved three criteria: 1) The stenotic occlusion of the terminal ICA and the proximal portion of anterior and/or middle cerebral arteries, 2) Abnormal vascular network evidenced in the arterial phase in proximity to the occlusion, and 3) Bilateral involvement.6 The clinical presentation of MMD is variable depending on the age of the patient which include: transient ischemic attacks, ischemic stroke, hemorrhagic stroke, headache, and cognitive dysfunction.40

Bibliometric analysis is a detailed statistical tool that has been widely used to review scientific research in various fields.12(34)38(33) Recently, bibliometric analyses, in the form of citations count, have been used as indicators of performance in the scientific literature.1) Bibliometric evaluation of neurosurgical publications has previously been published on pediatric neurosurgery, acoustic neuroma, intracerebral hemorrhage and meningioma.23(33) To the best of our knowledge, however, a bibliometric analysis has never been performed on MMD.

As bibliometric analyses have been conducted to assess the intellectual work in the academic fields, a comprehensive bibliometric analysis is required to assess the current publication trends on MMD. They help identify the metrics, trends, as well as the patterns of publications. This will enable the readership of the journals to gain evidence-based knowledge in the field of MMD. In the current study, we aim to conduct a bibliometric analysis to evaluate and assess the influence of the published literature in Moyamoya disease/syndrome by examining the most-cited articles in the literature.

MATERIALS AND METHODS

This bibliometric review study on the most-cited articles on MMD was performed using the Scopus database. A non-time restricted keyword-based search was performed in June 2020. The search keywords included the following keywords: “Moyamoya”, “Moyamoya disease”, and “Moyamoya syndrome”. The identified articles were ranked from highest to lowest based on their citation count and the most-cited top 100 articles were chosen for further examination. The citation per year (CY) was used to minimize time-bias, as some articles were recently published without having enough time to accumulate citations over the years. In reviewing the top-100 articles, we collected pertinent information to our review which included thirteen parameters.

The article-based parameters (title, year of publication, country of origin, contributing authors, speciality of the most contributing authors, contributing institutes, and publishing journal), and bibliometric-based parameters (citation count, citation per year, author’s H-index, Journal’s Source Normalized Impact Per Paper [SNIP], Journal’s SCImago Journal Rank [SJR], and the Journals Impact Factor [IF]). The categorization of the top-100 articles, after analyzing the studied entities in the most-cited articles, has classified the studies into seven categories. This included; Epidemiological studies, pathophysiological studies, clinical studies, clini-
coradiological studies, radiological studies, guidelines studies, and surgical management studies. Based on the identified journals of contribution, a quantified method was used to rank the most contributing journals based on the number of publications per journal. A journal with five or more publications from the top 100 list were considered to be highly contributing to the most-cited articles on MMD.

### RESULTS

The Scopus based search has identified 3,543 published articles on Moyamoya disease between 1955 and 2020. The Top-100 articles were published between 1977 and 2016 (Table 1). The top-100 received a total of 16,119 citations and 161.19 citations per year, with a 7.23% rate of self-citation for all contributing authors. Studying the

| Rank | Authors | Title | Journal Name | CC | CY |
|------|---------|-------|--------------|----|----|
| 1st  | Scott RM and Smith ER, 2009 | Moyamoya disease and moyamoya syndrome | New England Journal of Medicine | 763 | 69.36 |
| 2nd  | Suzuki J and Kodama N, 1983 | Moyamoya disease-A review | Stroke | 590 | 15.95 |
| 3rd  | Kuroda S and Houkin K, 2008 | Moyamoya disease: current concepts and future perspectives | The Lancet Neurology | 508 | 42.2 |
| 4th  | Fukui M, 1997 | Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis ('Moyamoya' disease) | Clinical Neurology and Neurosurgery | 502 | 21.8 |
| 5th  | [No author name available], 2012 | Guidelines for diagnosis and treatment of moyamoya disease (spontaneous occlusion of the circle of Willis): Research Committee on the Pathology and Treatment of Spontaneous Occlusion of the Circle of Willis; Health Labour Sciences Research Grant for Research on Measures for intractable Diseases | Neurologia Medico-Chirurgica | 348 | 43.5 |
| 6th  | Scott RM et al., 2004 | Long-term outcome in children with moyamoya syndrome after cranial revascularization by pial synangiosis | Journal of Neurosurgery | 337 | 21.06 |
| 7th  | Guo DC et al., 2009 | Mutations in smooth muscle alpha-actin (ACTA2) cause coronary artery disease, stroke, and moyamoya disease, along with thoracic aortic disease | American Journal of Human Genetics | 284 | 25.81 |
| 8th  | Kamada F et al., 2011 | A genome-wide association study identifies RNF213 as the first moyamoya disease gene | Journal of Human Genetics | 277 | 30 |
| 9th  | Karasawa J et al., 1978 | Treatment of moyamoya disease with STA-MCA anastomosis | Journal of Neurosurgery | 276 | 5.3 |
| 10th | Liu W., et al., 2011 | Identification of RNF213 as a susceptibility gene for moyamoya disease and its possible role in vascular development | PLoS ONE | 270 | 30 |
| 11th | Kuriyama S et al., 2008 | Prevalence and clinicopepidemiological features of moyamoya disease in Japan: Findings from a nationwide epidemiological survey | Stroke | 258 | 21.5 |
| 12th | Wakai K et al., 1997 | Epidemiological features of moyamoya disease in Japan: Findings from a nationwide survey | Clinical Neurology and Neurosurgery | 251 | 10.91 |
| 13th | Guzman R et al., 2009 | Clinical outcome after 450 revascularization procedures for moyamoya disease: Clinical article | Journal of Neurosurgery | 247 | 22.45 |
| 14th | Miyamoto S et al., 2014 | Effects of extracranial-intracranial bypass for patients with hemorrhagic moyamoya disease: Results of the Japan adult moyamoya trial | Stroke | 229 | 38.16 |
| 15th | Matsushima T et al., 1992 | Surgical treatment of moyamoya disease in pediatric patients—comparison between the results of indirect and direct revascularization procedures | Neurosurgery | 229 | 8.17 |
| 16th | Baba T et al., 2008 | Novel epidemiological features of moyamoya disease | Journal of Neurology, Neurosurgery and Psychiatry | 226 | 18.83 |
| 17th | Matsushima Y et al., 1981 | A new surgical treatment of moyamoya disease in children: A preliminary report | Surgical Neurology | 225 | 5.76 |

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| Rank | Authors                          | Title                                                                 | Journal Name                      | CC  | CY   |
|------|---------------------------------|----------------------------------------------------------------------|-----------------------------------|-----|------|
| 18th | Hallemeier CL et al., 2006      | Clinical features and outcome in North American adults with moyamoya phenomenon | Stroke                            | 219 | 15.64|
| 19th | Fukui M et al., 2000            | Moyamoya disease                                                      | Neuropathology                    | 219 | 10.95|
| 20th | Ikeda H et al., 1999            | Mapping of a familial moyamoya disease gene to chromosome 3p24.2-p26  | American Journal of Human Genetics | 218 | 10.38|
| 21st | Yamauchi T et al., 2000         | Linkage of familial moyamoya disease (spontaneous occlusion of the circle of Willis) to chromosome 17q25 | Stroke                            | 214 | 10.7 |
| 22nd | Chiu D et al., 1998             | Clinical features of moyamoya disease in the United States            | Stroke                            | 212 | 9.63 |
| 23rd | Uchino K et al., 2005           | Moyamoya disease in Washington State and California                   | Neurology                          | 205 | 13.66|
| 24th | Karasawa J et al., 1977         | A surgical treatment of "moyamoya" disease "Encephalo-Myo-synangiosis" | Neurologia medicochirurgica        | 198 | 4.6  |
| 25th | Dobson SR et al., 2002          | Moyamoya syndrome in childhood sickle cell disease: A predictive factor for recurrent cerebrovascular events | Blood                              | 189 | 10.5 |
| 26th | Fung LWE et al., 2005           | Revascularisation surgery for paediatric moyamoya: A review of the literature | Child’s Nervous System             | 186 | 12.4 |
| 27th | Yamashita M et al., 1983        | Histopathology of the brain vascular network in moyamoya disease      | Stroke                            | 185 | 5    |
| 28th | Karasawa J et al., 1992         | Long-term follow-up study after extracranial-intracranial bypass surgery for anterior circulation ischemia in childhood moyamoya disease | Journal of Neurosurgery            | 181 | 6.46 |
| 29th | Ullrich NJ et al., 2007         | Moyamoya following cranial irradiation for primary brain tumors in children | Neurology                          | 177 | 13.61|
| 30th | Inoue TK et al., 2000           | Linkage analysis of moyamoya disease on chromosome 6                   | Journal of Child Neurology         | 177 | 8.85 |
| 31st | Kuroda S et al., 2005           | Incidence and clinical features of disease progression in adult moyamoya disease | Stroke                            | 176 | 11.73|
| 32nd | Fukui M, 1997                   | Current state of study on moyamoya disease in Japan                    | Surgical Neurology                 | 172 | 7.47 |
| 33rd | Houkin K et al., 1996           | Surgical therapy for adult moyamoya disease: Can surgical revascularization prevent the recurrence of intracerebral hemorrhage? | Stroke                            | 163 | 6.79 |
| 34th | Ishikawa T et al., 1997         | Effects of surgical revascularization on outcome of patients with pediatric moyamoya disease | Stroke                            | 162 | 7.04 |
| 35th | Sakurai K et al., 2004          | A novel susceptibility locus for moyamoya disease on chromosome 8q23   | Journal of Human Genetics          | 148 | 9.25 |
| 36th | Miyatake S et al., 2012         | Homozygous c.14576G>A variant of RNF213 predicts early-onset and severe form of moyamoya disease | Neurology                          | 143 | 11   |
| 37th | Kuroda S et al., 2007           | Radiological findings, clinical course, and outcome in asymptomatic moyamoya disease: Results of multicenter survey in Japan | Stroke                            | 143 | 17.87|
| 38th | Kraemer M et al., 2008          | Moyamoya disease in Europeans                                         | Stroke                            | 141 | 11.75|
| 39th | Fujimura M et al., 2007         | Temporary neurologic deterioration due to cerebral hyperperfusion after superficial temporal artery-middle cerebral artery anastomosis in patients with adult-onset moyamoya disease | Surgical Neurology                 | 140 | 10.76|
| 40th | Yonekawa Y et al., 1997         | Moyamoya disease in Europe, past and present status                   | Clinical Neurology and Neurosurgery | 138 | 6    |
| 41st | Yoshida Y et al., 1999          | Clinical course, surgical management, and long-term outcome of moyamoya patients with rebleeding after an episode of intracerebral hemorrhage: An extensive follow-up study | Stroke                            | 136 | 6.47 |
| 42nd | Kawaguchi S et al., 2000        | Effect of direct arterial bypass on the prevention of future stroke in patients with the hemorrhagic variety of moyamoya disease | Journal of Neurosurgery            | 131 | 6.5  |
| 43rd | Imaizumi T et al., 1998         | Long-term outcomes of pediatric moyamoya disease monitored to adulthood | Pediatric Neurology                | 129 | 5.86 |

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| Rank | Authors | Title                                                                 | Journal Name                  | CC  | CY  |
|------|---------|-----------------------------------------------------------------------|-------------------------------|-----|-----|
| 44th | Ikezaki K et al., 1994 | Cerebral circulation and oxygen metabolism in childhood moyamoya disease: A perioperative positron emission tomography study | Journal of Neurosurgery       | 126 | 4.66|
| 45th | Kestle JRW et al., 1993 | Moyamoya phenomenon after radiation for optic glioma                  | Journal of Neurosurgery       | 126 | 4.84|
| 46th | Kobayashi E et al., 2000 | Long-term natural history of hemorrhagic moyamoya disease in 42 patients | Journal of Neurosurgery       | 124 | 6.2 |
| 47th | Kelly ME et al., 2006 | Progression of unilateral moyamoya disease: A clinical series         | Cerebrovascular Diseases      | 121 | 8.6 |
| 48th | Fujimura M et al., 2009 | Incidence and risk factors for symptomatic cerebral hyperperfusion after superficial temporal artery-middle cerebral artery anastomosis in patients with moyamoya disease | Surgical Neurology            | 120 | 5   |
| 49th | Kawaguchi S et al., 1996 | Characteristics of intracranial aneurysms associated with Moyamoya disease. A review of 111 cases | Acta Neurochirurgica          | 120 | 10.9|
| 50th | Duan L et al., 2012 | Moyamoya disease in China: Its clinical features and outcomes         | Stroke                        | 119 | 13.11|
| 51st | Zaharchuk G et al., 2011 | Arterial spin-labeling MRI can identify the presence and intensity of collateral perfusion in patients with moyamoya disease | Stroke                        | 118 | 13.11|
| 52nd | Okada Y et al., 1998 | Effectiveness of superficial temporal artery-middle cerebral artery anastomosis in adult moyamoya disease: Cerebral hemodynamics and clinical course in ischemic and hemorrhagic varieties | Stroke                        | 117 | 5.38|
| 53rd | Matsushima Y and Inaba Y, 1984 | Moyamoya disease in children and its surgical treatment. Introduction of a new surgical procedure and its follow-up angiograms | Child's Brain                 | 116 | 3.22|
| 54th | Kirugasa K et al., 1993 | Surgical treatment of moyamoya disease: Operative technique for encephalo-duro-arterio-myo-synangiosis, its follow-up, clinical results, and angiograms | Neurosurgery                   | 115 | 4.25|
| 55th | Kim SK et al., 2010 | Pediatric moyamoya disease: An analysis of 410 consecutive cases      | Annals of Neurology           | 114 | 11.4 |
| 56th | Ikezaki K et al., 1997 | A clinical comparison of definite Moyamoya disease between South Korea and Japan | Stroke                        | 112 | 4.86 |
| 57th | Kurokawa T et al., 1985 | Prognosis of occlusive disease of the circle of Willis (moyamoya disease) in children | Pediatric Neurology           | 111 | 3.17 |
| 58th | Mineharu Y et al., 2008 | Autosomal dominant moyamoya disease maps to chromosome 17q25.3.       | Neurology                     | 110 | 9.16 |
| 59th | Yoshimoto T et al., 1996 | Angiogenic factors in moyamoya disease                                | Stroke                        | 110 | 4.58 |
| 60th | Burke GM et al., 2009 | Moyamoya disease: A summary                                           | Neurosurgical Focus           | 109 | 9.9 |
| 61st | Goto Y and Yonekawa Y, 1992 | Worldwide distribution of moyamoya disease                            | Neurologia Medico-Chirurgica  | 109 | 3.89 |
| 62nd | Karasawa J et al., 1980 | Intracranial transplantation of the omentum for cerebrovascular moyamoya disease: A two-year follow-up study | Surgical Neurology            | 109 | 2.725|
| 63rd | Yamada I et al., 1995 | Moyamoya disease: Comparison of assessment with MR angiography and MR imaging versus conventional angiography | Radiology                     | 108 | 4.32 |
| 64th | Han DH et al., 2000 | A co-operative study: Clinical characteristics of 334 Korean patients with moyamoya disease treated at neurosurgical institutes (1976-1994) | Acta Neurochirurgica          | 107 | 5.35 |
| 65th | Choi JU et al., 1997 | Natural history of moyamoya disease: Comparison of activity of daily living in surgery and non surgery groups | Clinical Neurology and Neurosurgery | 105 | 4.56 |
| 66th | Kim SK et al., 2004 | Moyamoya disease among young patients: Its aggressive clinical course and the role of active surgical treatment | Neurosurgery                   | 104 | 6.5 |
| 67th | Golby AJ et al., 1999 | Direct and combined revascularization in pediatric moyamoya disease    | Neurosurgery                   | 104 | 4.95 |
| 68th | Lee M et al., 2009 | Quantitative hemodynamic studies in moyamoya disease A review         | Neurosurgical Focus           | 103 | 9.36 |
| 69th | Houkin K et al., 2000 | Neovascularization (angiogenesis) after revascularization in moyamoya disease. Which technique is most useful for moyamoya disease? | Acta Neurochirurgica          | 103 | 5.15 |

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| Rank | Authors                  | Title                                                                 | Journal Name                             | CC  | CY  |
|------|--------------------------|----------------------------------------------------------------------|------------------------------------------|-----|-----|
| 70th | Fujimura M et al., 2011  | Significance of focal cerebral hyperperfusion as a cause of transient neurologic deterioration after extracranial-intracranial bypass for moyamoya disease: Comparative study with non-moyamoya patients using n-isopropyl-p-[123I]iodoamphetamine single-photon emission computed tomography | Neurosurgery                             | 102 | 11.3|
| 71st | Ueki K et al., 1994      | Moyamoya disease: the disorder and surgical treatment                  | Mayo Clinic Proceedings                  | 102 | 3.92|
| 72nd | Kim JS, 2016             | Moyamoya disease: Epidemiology, clinical features, and diagnosis       | Journal of Stroke                         | 100 | 25  |
| 73rd | Khan N et al., 2003      | Moyamoya disease and Moyamoya syndrome: Experience in Europe; choice of revascularisation procedures | Acta Neurochirugica                        | 100 | 5.88|
| 74th | Takahashi A et al., 1993 | The cerebrospinal fluid in patients with moyamoya disease (spontaneous occlusion of the circle of Willis) contains high level of basic fibroblast growth factor | Neuroscience Letters                      | 98  | 3.62|
| 75th | Hoshimaru M et al., 1991 | Possible roles of basic fibroblast growth factor in the pathogenesis of moyamoya disease: An immunohistochemical study | Journal of Neurosurgery                   | 98  | 3.37|
| 76th | Kawaguchi T et al., 1996 | Multiple burr-hole operation for adult moyamoya disease               | Journal of Neurosurgery                   | 96  | 4   |
| 77th | Endo M et al., 1989      | Cranial burr hole for revascularization in moyamoya disease            | Journal of Neurosurgery                   | 95  | 2.87|
| 78th | Olds MV et al., 1987     | The surgical treatment of childhood moyamoya disease                   | Journal of Neurosurgery                   | 95  | 2.63|
| 79th | Miyamoto S et al., 1984  | Study of the posterior circulation in moyamoya disease. Clinical and neuroradiological evaluation | Journal of Neurosurgery                   | 95  | 3.06|
| 80th | Mineharu Y et al., 2006  | Inheritance pattern of familial moyamoya disease: Autosomal dominant mode and genomic imprinting | Journal of Neurology, Neurosurgery and Psychiatry | 94  | 5.87|
| 81st | Moroioka M et al., 2003  | Angiographic dilatation and branch extension of the anterior choroidal and posterior communicating arteries are predictors of hemorrhage in adult moyamoya patients | Stroke                                   | 94  | 6.17|
| 82nd | Houkin K et al., 1997    | Direct and indirect revascularization for moyamoya disease surgical techniques and peri-operative complications | Clinical Neurology and Neurosurgery       | 94  | 4.08|
| 83rd | Karasawa J et al., 1993  | Cerebral revascularization using omental transplantation for childhood moyamoya disease | Journal of Neurosurgery                   | 94  | 3.48|
| 84th | Mesiwala AH et al., 2008 | Long-term outcome of superficial temporal artery-middle cerebral artery bypass for patients with moyamoya disease in the US | Neurosurgical Focus                       | 92  | 7.66|
| 85th | Ryoo S et al., 2014      | High-resolution magnetic resonance wall imaging findings of moyamoya disease | Stroke                                   | 91  | 6.06|
| 86th | Jea A et al., 2005       | Moyamoya syndrome associated with down syndrome: Outcome after surgical revascularization | Pediatrics                                | 91  | 15.16|
| 87th | Houkin K et al., 1994    | Diagnosis of moyamoya disease with magnetic resonance angiography      | Stroke                                   | 91  | 3.5 |
| 88th | Miyamoto S, 2004         | Study design for a prospective randomized trial of extracranial-intracranial bypass surgery for adults with moyamoya disease and hemorrhagic onset: the Japan Adult Moyamoya Trial Group | Neurologia Medico-Chirurgica               | 89  | 5.56|
| 89th | Smith ER and Scott RM, 2005 | Surgical management of moyamoya syndrome                               | Skull Base                                | 88  | 3.66|
| 90th | Starke RM et al., 2009   | Clinical features, surgical treatment, and long-term outcome in adult patients with moyamoya disease: Clinical article | Journal of Neurosurgery                   | 87  | 7.9 |
| 91th | Mizoi K et al., 1996     | Indirect revascularization for moyamoya disease: Is there a beneficial effect for adult patients? | Surgical Neurology                        | 87  | 3.65|
| 92nd | Calamante F et al., 2001 | MR perfusion imaging in moyamoya syndrome: Potential implications for clinical evaluation of occlusive cerebrovascular disease | Stroke                                   | 86  | 4.52|
| 93rd | Schmit BP et al., 1996   | Acquired cerebral arteriovenous malformation in a child with moyamoya disease | Journal of Neurosurgery                   | 86  | 3.58|

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publications trends shows that the 1990s was the most active decade with 42 highly cited publications on MMD (Fig. 1). Studies on the surgical management and clinical outcome were the two most prevalent study categories (Fig. 2).

The most contributing country to the list was Japan by producing 60 articles from the list (Fig. 3). The Tohoku University School of Medicine was the most conducive institute to the list by contributing to 12 articles (Fig. 4). The Stroke Journal was the most active journal in terms of participation by publishing 23 articles from the top-100 list (Fig. 5). Houkin, K., a Japanese neurosurgeon, was the most prolific author to the top-100 list by authoring 15 articles from the list (Fig. 6). The number 1 article was authored by Scott R.M., and Smith E.R., which was published in the New England Journal of Medicine in 2009 entitled “Moyamoya disease and moyamoya syndrome”. The article received a total of 763 citations and 69.36 citation per year.

### DISCUSSION

The publications on Moyamoya disease in the late 70s focused on the surgical management. The authors targeted the effectiveness of anastomotic procedures (i.e., superficial temporal-middle cerebral artery STA-MCA), as a treatment modality for ischemic and/or neurolog-
ical deficits. Additionally, the first reported series on encephalo-my-o-synangiosis for patients with MMD was performed. In the 80s, however, the clinicoradiological aspect of MMD gained more interest, along with the surgical aspect. The angiographic findings of MMD were frequently correlated with clinical features of the disease. In the 90s, most publications were comparatively assessing direct vs. indirect revascularization techniques and the efficacy of STA-MCA vs. Encephalo-Duro-Arterio-Myo-Synangiosis (EDAMS) for the treatment of MMD. In the twenty-first century, MMD was being addressed from all aspects including the surgical, clinical, clinicoradiological, pathophysiological, radiological, epidemiological, and guideline-related.
In the top-100 list, the article with the highest citation count (CC) was published in the *New England Journal of Medicine* (2009) by Scott R.M. and Smith E.R. The article received a total of 763 CC and 69.3 citations per year (CY). The authors have comprehensively reviewed Moyamoya disease and syndrome from a demographic, pathogenetic, and treatment perspectives. In children and adults, MMD has been recognized to be the culprit of stroke. A prompt identification of the disease early in the course to facilitate therapy is essential to achieve excellent clinical outcome. In patients with MMD, treatment with revascularization surgery has been shown to be effective in the prevention of stroke.

The second highest cited article in our list was published in the *Stroke* (Journal) in 1983 by Suzuki J. and Kodama N, with 590 CC and 15.9 CY. The authors retrospectively evaluated 100 cases of MMD between
1961 and 1980. They have outlined the reason for the various clinical presentations in children vs. adults, emphasized on the angiographic findings of MMD, and its possible pathophysiology. Differences in the clinical features and radiological findings between children and adults were observed. In children, recurrent events of sudden hemiplegia have been commonly noted. In adults, however, intracranial hemorrhage was a common clinical presentation.

Surgical management

A quarter of the publications were targeting the surgical management of MMD. Fung et al. published an article (ranked 26th) with 186 CC and 12.4 CY. The authors performed a systematic review of the literature (1966–2004) discussing revascularization techniques and outcomes in children. Of note, 87% of the patients experienced symptomatic benefit, defined by complete resolution, reduction of the symptoms or remained asymptomatic, for a mean of approximately five years of follow-up postoperatively. Another article (ranked 14th) was published by Miyamoto and his colleagues with 229 CC and 38.1 CY. The authors conducted a prospective randomized controlled trial (JAM trial), it was concluded that the direct revascularization, in hemorrhagic Moyamoya, can improve the prognosis and lessen the rate of rebleeding after five years of follow-up, although the evidence is arguable.

To date, there are no well-settled recommendations of a particular type of revascularization over another. Direct techniques, first performed by Kikuchi and Karasawa among Moyamoya disease cases in 1973, have shown preferable prognosis outcomes but debatable rates of complications and longer operative time than indirect techniques. Karasawa et al. introduced revascularization in Moyamoya disease in 1980 with which it can be utilized as an option in cases where extracranial-intracranial anastomosis is difficult to perform. Another article (ranked 83rd) in 1993 which has received 94 CC and 3.4 CY. The authors investigated the application of omental transplantation to manage ischemic changes in the vascular territories of the anterior/middle/posterior arteries. Neurological improvement was noted in patients with omental transplantation. The authors recommended omental transplantation to manage ischemic changes in the anterior/posterior cerebral arteries territories.

Clinical studies

Articles in the clinical category, comprising 25% of
the top-100 list, peaked in 1985-2016. Kobayashi et al. published an article (ranked 46th) with 124 CC and 6.2 CY.22) The main aim of their study was to assess the long-term follow-up and natural history of hemorrhagic Moyamoya disease in 42 patients.22) In their cohort, the occurrence of rebleeding in hemorrhagic MMD was associated with worse prognosis.22) Clinically, MMD can present with different clinical manifestations including; stroke (hemorrhagic/ischemic), epilepsy, cognitive impairment, and/or headaches.21)32) Although it is largely influenced by geographical differences and age; ischemic presentation is common among the pediatric population.26) In adults, hemorrhagic presentation is more prevalent in east Asian countries.32) Yet, ischemic stroke is more prevalent in other populations.23) Additionally, it is worth noting that MMD can be isolated or present as a syndrome accompanying other conditions (e.g., sickle cell disease).11)28) The disease course, in both adults and pediatric age groups, is progressive and patients are likely to experience deteriorating neurological and cognitive functions, if left untreated.15)22)27)

Clinicoradiological studies
The clinicoradiological category gained popularity between the years of 1983 to 2012. Hallemeier et al. in their article (ranked 18th) concluded that North American patients with bilateral involvement of MMD and ischemic symptoms are rendered susceptible to recurrent strokes.10) On the other hand, Kuroda et al. published an article (ranked 37th) with 143 CC and 17.8 CY.26) The authors conducted a multi-center study to investigate the clinical and radiological features of asymptomatic MMD.26) Their findings demonstrated that asymptomatic MMD could potentially cause lethal neurological sequelae such as ischemic and hemorrhagic strokes.26) Consequently, such patients require a periodic clinical assessment to further delineate their long-term outcome.26)

Pathophysiological studies
Articles addressing the pathogenesis of MMD were published between 1981-2012. The key area of interest was on the histopathological features, molecular aspect, and genetic/inheritance patterns of MMD. The highly characteristic patterns of the disease has led the investigations of underlying genetic association to start in the late 1990s.13) Proportion of 6% and 10%, in the US and Japanese population of Moyamoya disease affected individuals, are found to have a 1st degree family relative with the same condition.5307) RNF213 gene, non-exclusively, found to be attributed to the susceptibility of its development in some Asian populations.16)28) Guo et al. published an article (ranked 7th) with 284 CC and 25.8 CY.9) The authors investigated possible mutations contributing to the development of MMD.9) Their study findings revealed that single-gene mutations can cause diffuse vascular diseases that may play a central role in the pathogenesis of MMD.9) Similarly, Kamada et al. published an article (ranked 8th) with 277 CC and 30 CY. The authors performed a genome-wide analysis of 72 patients with MMD and subsequently compared them to 45 patients in the control group.16) They have demonstrated that RNF213 gene serves as the first gene implicated in the pathogenesis of MMD.16)

Radiological studies
The majority of publications in the radiological category peaked in 1994 to 2014. The key topics during that era were utility and efficacy of magnetic resonance imaging (MRI), digital subtraction angiography (DSA), and positron emission tomography (PET). Ikezaki et al. published an article (ranked 44th) with 126 CC and 4.6 CY.14) The authors conducted a peri-operative study to investigate the role of positron emission tomography (PET) scan in oxygen metabolism of pediatric MMD patients.14) PET scans were essential to investigate cerebral metabolism and circulation in the evaluation of pediatric patients with MMD.14) Contrarily, Yamada et al. published an article (ranked 63rd) with 108 CC and 4.3 CY.42) The authors evaluated the role of MRI and magnetic resonance angiography (MRA) in describing MMD.42) Their study findings revealed that the combination of MRI and MRA yields excellent results in the identification of steno-occlusive disease and moyamoya vessels.42)
Epidemiological studies

Articles investigating the epidemiological perspective of MMD peaked in publication between 1992-2008, with most studies being conducted in Japan. Kuriyama et al. conducted a questionnaire-based survey, (ranked 11th) with 258 CC and 21.5 CY, to estimate the prevalence of MMD. They have proposed that the prevalence rate of MMD in Japan has escalated in recent years, due to the diagnosis of newly confirmed cases. In the article of Goto and Yonekawa (ranked 61st) published in 1992 with 109 CC and 3.8 CY, the worldwide geographical distribution of MMD was commonly observed in Asia and non-Caucasian populations.

Guidelines

Two articles comprehensively investigated the guidelines implicated in the diagnosis and management of MMD. The first guideline article was published in 1997 with 502 CC and 21.8 CY. The second article was published in 2012, to which it received 348 CC and 43.5 CY. Interestingly, the two articles ranked 4th and 5th in the top-100 list of the present study, although they were published 15 years apart.

Limitations

This bibliometric analysis is not free from inherent limitations. The citation received for old citation over-time can lead to citation over representation and this time-bias can be minimized by using the citation per year which was used in our review. The fact that citation-based analysis of articles does not represent the scientific value of the article and the proper use of bibliometrics with the conjunction of peer reviewed articles will provide a holistic view in any studied entity. The Scopus database has some limitations in which they are improving over time. The detailed citation coverage for articles published before 1970 is not included in their bibliometric tools which can under represent the citation count for articles published prior to that time. Additionally, the rate of self-citation can insignificantly inflate the citations of articles. However, self-citation was minor in our bibliometric review on MMD.

CONCLUSIONS

The majority of publications investigating Moyamoya disease were targeting the surgical and clinical aspects. In the management of MMD, the effectiveness of anastomotic procedures has gained acceptance, especially in the treatment of ischemic and neurological deficits. The top two most cited articles were published, as reviews, in the New England Journal of Medicine and Stroke. Neurosurgeons were contributing to most of the publications in the top-100 list. In the 21st century, authors were investigating and publishing articles on MMD from all clinical perspectives. The present bibliometric analysis highlighted the most reputable key articles to researchers in the field of Moyamoya disease.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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