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Original Article

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.[43)]

Bibliometric analysis is a detailed statistical tool that has been widely used to review scientific research in various fields.[12)34)38)] 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.[2)3)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

Table 1. The top-100 most cited articles on Moyamoya disease ranked by their corresponding citation count

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 clinicoepidemiological 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 medico-chirurgica	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	Kinugasa 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 Neurochirurgica	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	Morioka 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
91st	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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Rank	Authors	Title	Journal Name	CC	CY
94th	Yamada I et al., 1992	Moyamoya disease: Diagnosis with three-dimensional time-of-flight MR angiography	Radiology	86	3.071
95th	Maeda M and Tsuchida C, 1999	'Ivy sign' on fluid-attenuated inversion-recovery images in childhood moyamoya disease	American Journal of Neuroradiology	85	4.04
96th	Iwama T et al., 1996	The relevance of hemodynamic factors to perioperative ischemic complications in childhood moyamoya disease	Neurosurgery	83	3.45
97th	Oka K et al., 1981	Cerebral haemorrhage in moyamoya disease at autopsy	Virchows Archiv A Pathological Anatomy and Histology	83	2.12
98th	Houkin K et al., 2005	Novel magnetic resonance angiography stage grading for moyamoya disease	Cerebrovascular Diseases	82	4.31
99th	Yilmaz EY et al., 2001	Moyamoya: Indiana University Medical Center experience	Archives of Neurology	82	5.46
100th	Uchino H et al., 2012	Predictors and clinical features of postoperative hyperperfusion after surgical revascularization for moyamoya disease: A serial single photon emission CT/positron emission tomography study	Stroke	81	10.12

CC, citation count; CY, citation per year; MRI, magnetic resonance imaging

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-

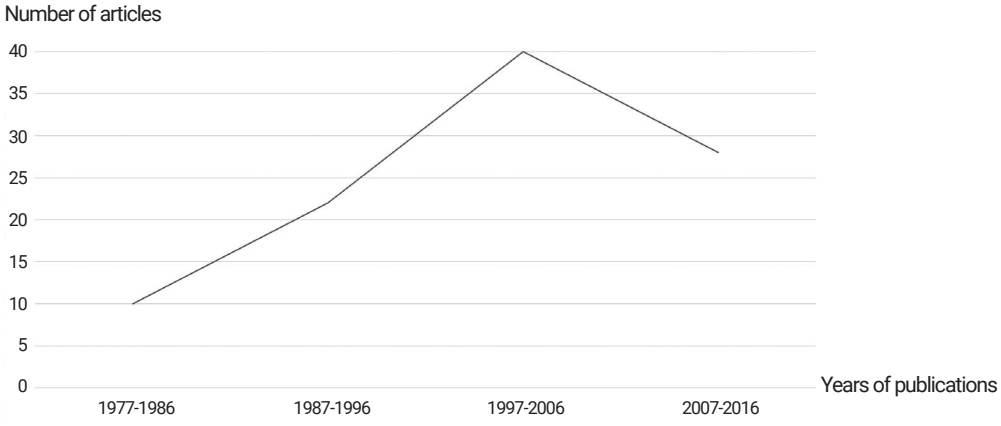

Fig. 1. Publication trends of the top-100 most cited articles on Moyamoya disease between 1977-2016.

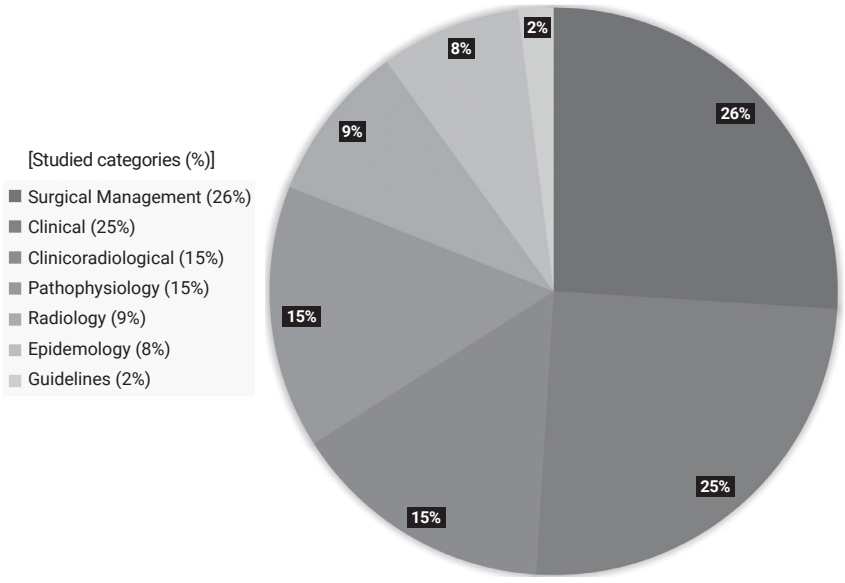

Fig. 2. Categorical distribution of Moyamoya disease in the top-100 cited articles.

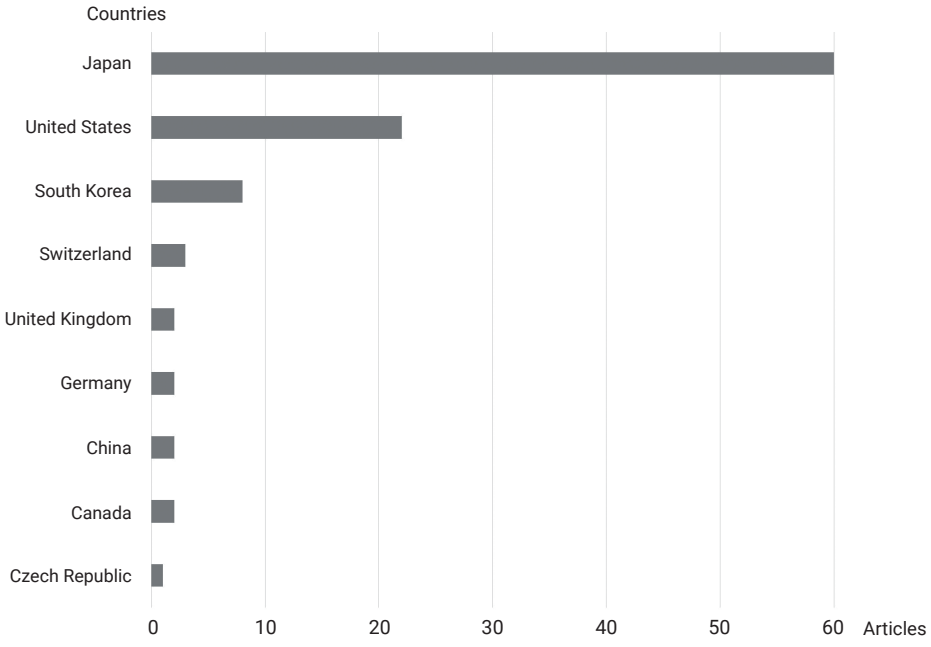

Fig. 3. The country of origin of the articles on Moyamoya disease in the top-100 cited articles.

ical deficits. Additionally, the first reported series on encephalo-myo-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.

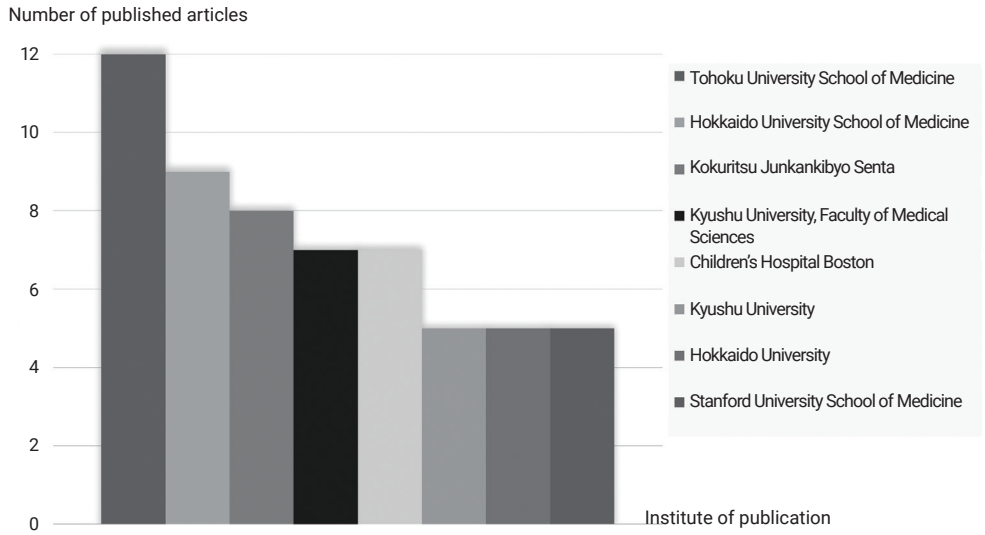

Fig. 4. Affiliated institutes contributing to the highest cited articles on Moyamoya disease.

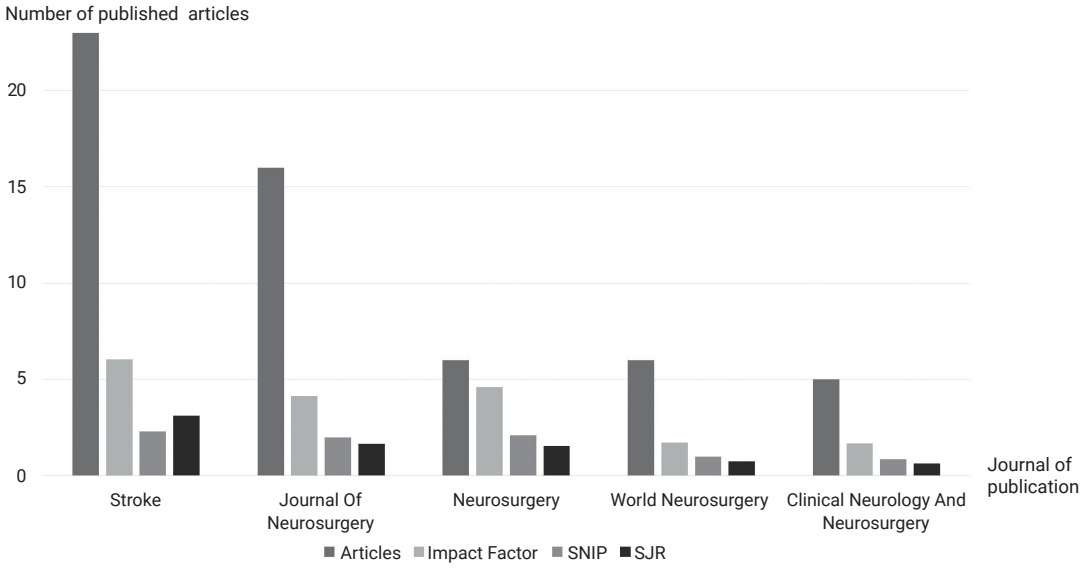

Fig. 5. The top journals publishing articles on Moyamoya disease and their corresponding impact factor, Journal's SNIP, Journal's SJR. SNIP, Source Normalized Impact Per Paper; SJR, Journal's SCImago Journal Rank.

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.[36] The article received a total of 763 CC and 69.3 citations per year (CY).[36] The authors have comprehensively reviewed Moyamoya disease and syndrome from a demographic, pathogenetic, and treatment perspectives.[36] In children and adults, MMD has been recognized to be the culprit of stroke.[36] A prompt identification of the disease early in the course to facilitate therapy is essential to achieve excellent clinical outcome.[36] In patients with MMD, treatment with revascularization surgery has been shown to be effective in the prevention of stroke.[36]

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.[39] The authors retrospectively evaluated 100 cases of MMD between

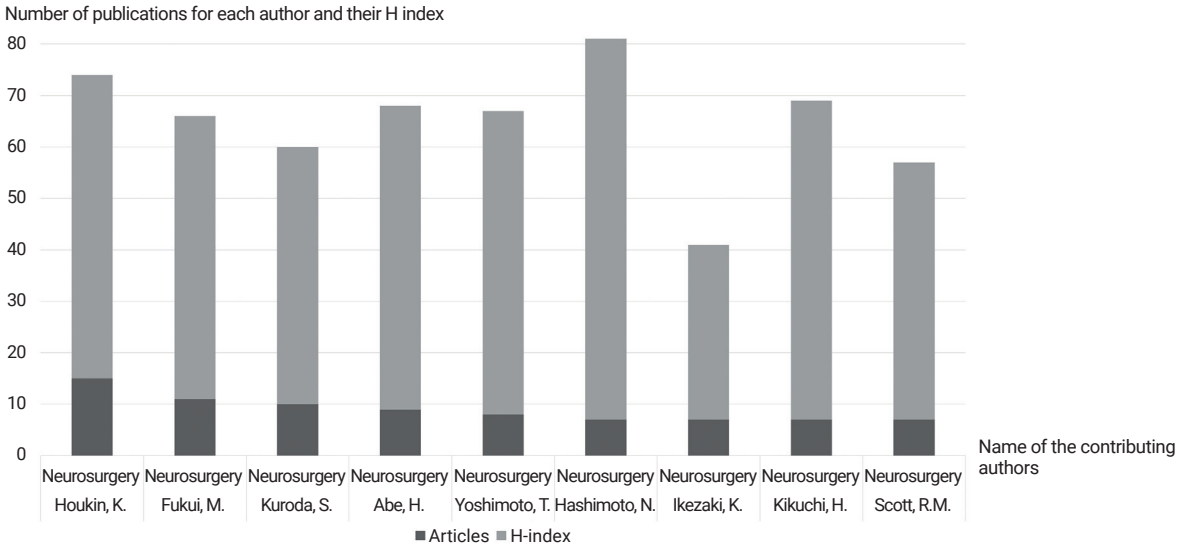

Fig. 6. Affiliated institutes contributing to the highest cited articles on Moyamoya disease.

1961 and 1980.[39] 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.[39] Differences in the clinical features and radiological findings between children and adults were observed.[39] In children, recurrent events of sudden hemiplegia have been commonly noted. In adults, however, intracranial hemorrhage was a common clinical presentation.[39]

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.[7] The authors have performed a systematic review of the literature (1966-2004) discussing revascularization techniques and outcomes in children.[7] 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.[7] Another article (ranked 14th) was published by Miyamoto and his colleagues with 229 CC and 38.1 CY.[31] The authors conducted a prospective randomized controlled trial to investigate the feasibility of extracranial-intracranial bypass in reducing the incidence of rebleeding.[31] In their 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.[4][31]

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.[5][19][30] 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.[17][30] Karasawa et al. published their article (ranked 83rd) in 1993 which has received 94 CC and 3.4 CY.[18] The authors investigated the application of omental transplantation to manage ischemic changes in the vascular territories of the anterior/middle/posterior arteries.[18] Neurological improvement was noted in patients with omental transplantation.[18] The authors recommended omental transplantation to manage ischemic changes in the anterior/posterior cerebral arteries territories.[18]

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.[28] 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.[6][37] *RNF213* gene, non-exclusively, found to be attributed to the susceptibility of its development in some Asian populations.[16][29] 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.[25] They have proposed that the prevalence rate of MMD in Japan has escalated in recent years, due to the diagnosis of newly confirmed cases.[25] 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.[8]

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 overtime 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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