1-1-2016

Arteriovenous malformations of the corpus callosum: Pooled analysis and systematic review of literature.

Aqueel H. Pabaney
Henry Ford Health System

Rushna Ali
Henry Ford Health System, RAli1@hfhs.org

Maximillian K. Kole
Henry Ford Health System, mkole1@hfhs.org

Ghaus Malik
Henry Ford Health System, gmalik1@hfhs.org

Recommended Citation

Pabaney AH, Ali R, Kole M, and Malik GM. Arteriovenous malformations of the corpus callosum: Pooled analysis and systematic review of literature. Surg Neurol Int 2016; 7(Suppl 9):S228-236.

SNI
SURGICAL NEUROLOGY INTERNATIONAL
SNI: Cerebrovascular, a supplement to Surgical Neurology International
OPEN ACCESS
For entire Editorial Board visit : http://www.surgicalneurologyint.com
Editor: James I. Ausman, MD, PhD University of California, Los Angeles, CA, USA

Arteriovenous malformations of the corpus callosum: Pooled analysis and systematic review of literature

Aqueel H. Pabaney†, Rushna Ali†, Maximillian Kole, Ghaus M. Malik

Department of Neurological Surgery, Henry Ford Hospital, 2799 W. Grand Blvd, Detroit, MI 48202, USA

E-mail: *Aqueel H. Pabaney - apabane1@hfhs.org; Rushna Ali - rali1@hfhs.org; Maximillian Kole - mkole1@hfhs.org; Ghaus M. Malik - gmalik1@hfhs.org
*Corresponding author

†These authors contributed equally to the manuscript.

Received: 24 October 15 Accepted: 02 February 16 Published: 01 April 16

Abstract

Background: Arteriovenous malformations (AVMs) of the corpus callosum (CC) are rare entities. We performed a systematic review of the available literature to better define the natural history, patient characteristics, and treatment options for these lesions.

Methods: A MEDLINE, Google Scholar, and The Cochrane Library search were performed for studies published through June 2015. Data from all eligible studies were used to examine epidemiology, natural history, clinical features, treatment strategies, and outcomes of patients with CC-AVMs. A systematic review and pooled analysis of the literature were performed.

Results: Our search yielded 37 reports and 230 patients. Mean age at presentation was 26.8 years (±13.12 years). AVMs were most commonly located in the splenium (43%), followed by the body (31%), and then the genu (23%) of the CC. A Spetzler-Martin grade of III was the most common (37%). One hundred eighty-seven (81.3%) patients presented with hemorrhage, 91 (40%) underwent microsurgical excision, and 87 (38%) underwent endovascular embolization. Radiosurgery was performed on 57 (25%) patients. Complete obliteration of the AVM was achieved in 102 (48.1%) patients and approximately twice as often when microsurgery was performed alone or in combination with other treatment modalities (94% vs. 49%; $P < 0.001$). Mean modified Rankin Scale (mRS) at presentation was 1.54 and mean mRS at last follow-up was 1.31. This difference was not statistically significant ($P = 0.35$).

Conclusion: We present an analysis of the pooled data in the form of a systematic review focusing on management of CC-AVMs. This review aims to provide a valuable tool to aid in decision making when dealing with this particular subtype of AVM.

Key Words: Arteriovenous malformation, corpus callosum, endovascular therapy

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/2152-7806.179579
Quick Response Code:

INTRODUCTION

Arteriovenous malformations (AVMs) of the corpus callosum (CC) are distinct clinical as well as surgical entities. They are known to cause recurrent hemorrhage more frequently as compared to more superficial, pial AVMs.[18] Optimal treatment of these AVMs is controversial not only because they tend to cause mild

How to cite this article: Pabaney AH, Ali R, Kole M, Malik GM. Arteriovenous malformations of the corpus callosum: Pooled analysis and systematic review of literature. Surg Neurol Int 2016;7:S228-36.
http://surgicalneurologyint.com/Arteriovenous-malformations-of-the-corpus-callosum:-Pooled-analysis-and-systematic-review-of-literature/

clinical impairment in the absence of major hemorrhage but also because surgical resection may result in permanent neurologic deficit as well neuropsychological deterioration. In 1951, Basset reported the first successful surgical resection of a CC-AVM.[7] Since surgical resection can be technically challenging, less invasive therapies have been developed recently for the treatment of these lesions. Endovascular treatment has occasionally been employed; however, complete obliteration is rarely achieved and aggressive embolization can lead to major neurological deterioration and possibly treatment-related death. Stereotactic radiosurgery (RS) is a safe and effective treatment for select patients with CC-AVMs as it reduces the risk of damage to critical adjacent vascular structures. Even large, ruptured AVMs can be treated successfully with minimal morbidity and a low risk of repeated hemorrhage.[29] Given the paucity of literature on CC-AVMs, the authors aim to delineate the natural history, vascular anatomy, treatment approaches, and outcomes of this uncommon vascular anomaly.

METHODS

We undertook a review of the literature on CC-AVMs. Our methods were in accordance with the systematic review section of the preferred reporting items for systematic reviews and meta-analyses statement.[33] Subsequently, a pooled analysis of the literature was performed.

Search strategy

Two independent reviewers (Rushna Ali and Aqueel H. Pabaney) performed a comprehensive electronic search for articles on CC-AVMs. The search included MEDLINE (PubMed and Ovid), Google Scholar, and The Cochrane Library from January 1970 to June 2015. These databases were queried using keywords and MeSH terms "arteriovenous malformation," "AVM," "corpus callosum," "radiosurgery," "endovascular therapy," "microsurgery," "surgical resection," and "outcome." A full-text version was obtained for all studies the reviewers considered relevant. To identify additional resources, we manually searched references of articles that could be potentially relevant. No restrictions were imposed based on publication dates, types, or language.

Study selection

We selected articles reporting clinical outcomes for patients with CC-AVMs treated with any of the three treatment modalities (i.e., microsurgery, endovascular embolization, and RS). We included articles that documented patient age, clinical presentation, location of AVM, treatment rendered, and outcomes (modified Rankin Scale [mRS] or posttreatment complications). We included studies not published in English if they met the above selection criteria. We excluded articles that did not report the location of AVM, treatment modality, or outcomes as well as abstracts and letters to the editor that were not published as full reports. Disagreements between the two reviewers about the relevance of a particular study were resolved by discussion and consensus with a third reviewer (Ghaus M. Malik or Maximillian Kole).

Data extraction

Two independent reviewers (Rushna Ali and Aqueel H. Pabaney) performed data extraction. Discrepancies were settled by discussion and consensus. Information was gathered from eligible articles (systematic review) using data abstraction forms, which included the following information: Total number of patients, patients' age and gender, location of the AVM within the CC as defined by Yasargil's classification,[52] clinical presentation, treatment strategy, obliteration rates, and outcome measures.

Definition of variables

AVM location was divided into those present in the genu, body, or splenium of the CC. Presenting symptoms were categorized as hemorrhage (i.e., subarachnoid [SAH], intraventricular, and intracerebral [ICH]), seizures, headaches, and neurological deficits. Others were classified as incidental findings. Treatment strategy was divided into microsurgical resection, endovascular embolization, radiation therapy, or a combination of two or more strategies. Obliteration rates were defined as either complete obliteration or residual AVM. Long-term outcome was defined by mRS or postoperative complications.

Statistical analysis

We used an independent two-tailed *t*-test (Welch generalization of the Student's *t*-test, Microsoft Excel, 2013, Redmond, WA, USA) to compare preoperative and postoperative mRSs. Chi-square test was utilized to compare the difference in AVM obliteration rates achieved by various treatment modalities. *P* value of 0.05 was considered statistically significant.

RESULTS

Eligibility criteria described in the methods section were met by 37 articles, with a total of 230 patients [Table 1]. Individual patient data could be obtained from 31 reports. All reports were retrospective observational cohorts, case reports, and case series. No prospective cohorts or randomized studies were found.[2-6,8,10-12,15-21,23-34,37-42,47,48,50,53,54]

The mean patient age at presentation was 26.82 years (±13.12 years; range 2–61 years; median 25 years) with a slight male (55%) preponderance. CC-AVMs were most commonly located in the splenium (99 patients; 43%), followed by the body (72 patients; 31%), and then the genu (54 patients; 23%). Two or more areas of CC were

Table 1: Studies reporting patients with corpus callosum arteriovenous malformations

Author and year	Number of patients	Mean age (years)	Male: female	Presentation	Location of AVM	S-M grade	Treatment	Obliteration status*	Mean initial mRS	Mean follow-up mRS	Complications
Akimoto 2003[2]	1	27	0:1	HA, numbness	Splenium, body	IV	Surgery	Complete	1	0	None
Andreussi 1978[4]	1	13	0:1	HA, LOC	Genu	II	Surgery	Complete	5	1	None
Baiz 1964[5]	1	16	0:1	HA, blurred vision	Splenium, body	IV	Surgery	Complete	4	0	Foot weakness
Bartal 1970[6]	1	7	1:0	Hemiplegia	Genu, body	III	Surgery	Complete	5	3	None
Bendavid 2004[8]	1	2	0:1	HA	Genu	III	Surgery	Complete	1	0	None
Buklina 2002[10]	36	25	NA	Hemorrhage	Genu: 9 Body: 7 Splenium: 20	NA	Surgery: 29 Observed: 7	NA	NA	NA	NA
Castro-Caldas 1989[11]	1	30	0:1	HA	Body, splenium	IV	Surgery	Complete	1	0	None
Cone 1979[12]	3	25	2:1	HA, seizures, IVH	Genu: 1 Splenium: 2	II: 1 V: 1 NA: 1	Surgery: 1 Observed: 2	Complete	0	0	None
Da Pian 1980[13]	2	25.5	1:1	SAH	Body: 1 Genu: 1	II: 1 III: 1	Surgery	Complete	4.5	2	None
Dikel *et al*., 2001[15]	1	6	1:0	HA	Holocallosal	IV	Surgery + embolization + XRT	NA	1	0	None
Ganapathy *et al*., 2003[16]	1	39	1:0	HA	Holocallosal	V	XRT	Complete	1	0	None
Garza-Mercado *et al*., 1987[17]	1	15	0:1	SAH	Holocallosal	IV	Surgery	NA	4	6	Death
Guidetti 1982[18]	15	35	10:5	SAH: 14 Deficit: 1	Body: 3 Genu: 2 Splenium: 10	NA	Surgery: 11; Observed: 4	Complete: 9 NA: 3	1.87	2.87	Dead: 1 Hemianopsia: 3 Hemiparesis: 1 Memory loss: 1
Herzig 2000[19]	1	45	0:1	SAH	Splenium	III	XRT	Complete	0	0	None
Houtteville 1989[20]	1	18	0:1	SAH	Body	II	XRT	NA	0	1	None
Juhasz 1978[21]	2	14	0:2	HA	Splenium	III	Surgery	Complete	0	1	None
Kohmura 1990[23]	1	23	1:0	Incidental	Splenium	III	Surgery	Incomplete	0	1	None
Kosary 1978[24]	3	20	2:1	HA	Splenium	II	Surgery: 3	Complete: 2 NA: 1	2	3	Hemianopsia: 1 Death: 1
Koyanagi 1985[25]	1	NA	1:0	NA	Body	III	Embolization + surgery	Complete	NA	NA	NA
Kunc 1974[26]	8	NA	NA	SAH	Genu: 4 Body: 1 Splenium: 2 Holocallosal: 1	NA	Surgery, 4	NA	NA	Excellent: 3	None
Lobato 2002[27]	1	58	0:1	SAH	Body	IV	Embolization + surgery	Complete	0	0	Memory deficit
Machado 1984[28]	1	25	0:1	SAH	Body and splenium	IV	Surgery	Complete	0	1	None

Contd..

Table 1: Contd...

Author and year	Number of patients	Mean age (years)	Male: female	Presentation	Location of AVM	S-M grade	Treatment	Obliteration status*	Mean initial mRS	Mean follow-up mRS	Complications
Maruyama 2005[29]	32	25	15:17	SAH: 28 Other: 4	Genu: 5 Body: 9 Splenium: 19	I: 2 II: 8 III: 17 IV: 4 V: 1	XRT	Complete: 21 Incomplete: 11	NA	NA	Dysarthria: 1
McDonald 2001[30]	1	47	1:0	Memory impairment	Splenium	III	Embolization + XRT	NA: 1	0	0	Memory impairment
Milhorat 1970[31]	1	16	1:0	SAH	Body	IV	Surgery	Complete	0	0	None
Mohanty 2011[32]	1	13	1:0	SAH	Body	II	XRT	Complete	0	0	None
Orozco 2013[34]	2	48.5	1:1	SAH and seizure	Body	III and IV	Embolization: 1 Embolization + surgery: 1	Complete in 1 patient undergoing surgery Incomplete: 1	3	2.5	Hemiparesis
Picard 1996[37]	43	30	25:18	SAH: 36 Seizures: 4 HA: 2 Ataxia: 1	Genu: 11 Body: 6 Splenium: 20 Holocallosal: 9	NA	Embolization: 43 XRT: 9	Complete: 8 (4 embolization, 4 embolization + XRT) Incomplete: 35	NA NA	NA	Death: 1 Hemiparesis: 1
Robert 2015[39]	38	31	17:21	Hemorrhage: 30; Incidental: 1; Seizures: 4; Neuro deficit: 3	Genu: 14 Body: 19 Holocallosal: 5	I: 2 II: 20 III: 9 IV: 6 V: 1	Embolization: 38 XRT: 9	Complete: 22 (16 embolization, 6 embolization + XRT) Incomplete: 10	NA	1.05	Neuro deficit NOS: 5
Sell 1997[40]	1	20	0:1	SAH	Fornix and splenium	III	Surgery	Incomplete	5	2	Memory deficits
Shi 1987[41]	5	25	4:1	SAH	Body	II: 1 III: 4	Surgery	Complete	2	0.8	Seizures; 1
Shimizu 2001[42]	1	50	1:0	Incidental	Body	III	XRT + embolization	Complete	1	0	None
Uchino 1989[47]	1	16	0:1	SAH	Genu and body	V	XRT	Incomplete	2	1	None
Valenstein 1987[48]	1	39	1:0	SAH	Splenium	II	Surgery	Complete	2	2	Memory deficits
Wang 2001[50]	1	51	1:0	SAH	Genu and body	V	Surgery	Complete	5	4	Hemiparesis
Yasargil 1976[54]	8	23.6	5:3	SAH	Genu: 5 Body: 6 Holocallosal: 1	III: 1 IV: 4 V: 3	Surgery	Complete	1.25	1.1	Hemiparesis: 1
Yasargil 1976[53]	10	28.2	8:2	SAH: 7 HA: 3	Splenium: 10	III: 6 IV: 2 V: 2	Surgery	Complete: 5 NA: 5	0.6	0.7	Hemianopsia: 3 Seizures: 2

*Angiographic obliteration in patients undergoing any treatment modality. NA: Data not available, LOC: Loss of consciousness, HA: Headache, IVH: Intraventricular hemorrhage, SAH: Subarachnoid hemorrhage, ICH: Intracranial hemorrhage, XRT: Radiation therapy, S-M: Spetzler-Martin, AVM: Arteriovenous malformation, NOS: Not otherwise specified

involved in 13 patients (6%) whereas the entire CC was involved in 21 (9%). Spetzler-Martin grade was reported or could be calculated for 135 patients. The most common Spetzler-Martin grade was 3 (51 patients; 37%) followed by 2 (38 patients; 28%).

Overall, analysis of 230 patients showed that 187 (81.3%) presented with hemorrhage, 5 with focal neurological deficit, 10 with seizures without hemorrhage, and 18 with headaches as the primary symptom. Ninety-one (40%) patients underwent microsurgical resection of the AVM. Endovascular embolization was performed either as a preoperative adjunct or as a stand-alone treatment modality in 87 (38%) patients. RS was administered to 57 (25%) patients while two or more treatment modalities

were used in combination in 23 (10%). Cumulatively, 212 (92.1%) patients received treatment whereas 18 (7.8%) were managed conservatively. Complete obliteration of the AVM was achieved in 102 (48.1%) patients. This was accomplished by microsurgery in 44 (43.1%) patients, embolization in 20 (19.6%), RS in 24 (23.5%), and combination therapy in 14 (13.7%).

Residual AVM was observed in 60 (28.3%) patients. Half of these patients underwent stand-alone embolization (30 patients; 50%), 12 (20%) RS alone, and 15 (25%) received combination therapy of embolization and RS. Of those who underwent surgical resection, only 3 (5%) patients had a residual nidus. Stand-alone surgical resection or in combination with other treatment modalities resulted in complete obliteration in 47 of 50 (94%) patients whereas only 55 (49%) of 112 who underwent embolization alone, RS alone, or combination therapy with embolization and RS achieved complete obliteration. This difference was found to be statistically significant ($P < 0.001$). Thirty-two patients encountered complications in the immediate posttreatment period; however, only 26 patients were left with long-lasting adverse effects from treatment. Three deaths were noted in the literature, 2 occurred posttreatment, and 1 patient died after managed conservatively. Mean mRS at presentation was 1.54 and mean mRS at last follow-up was 1.31. The difference between the initial and final mRS was not statistically significant ($P = 0.35$) [Table 2].

Table 2: Patient characteristics with corpus callosum arteriovenous malformations

Patient characteristics	*n*
Number of reports	37
Eligible patients	230
Mean age (±SD) years	26.8 (±13.12)
Gender (*n*=185) (%)	
Males	101 (54.5)
Females	84 (45.4)
Presentation (%)	
Hemorrhage	187 (81.3)
Seizures	10 (4)
Headache	18 (7.8)
Others	15 (6.5)
Location (%)	
Genu	54 (23.4)
Body	72 (31.3)
Splenium	99 (43)
>Two locations	13 (5)
Holocallosal	21 (9.1)
S-M grade (*n*=135) (%)	
I	4 (2.9)
II	38 (28)
III	51 (37.7)
IV	27 (20)
V	15 (11.1)
Treatment (%)	
Surgery	91 (39.5)
Embolization	87 (37.8)
Radiation treatment	57 (24.7)
Combination therapy	23 (10)
Complications (%)	
Immediate	32 (13.9)
Long-term	26 (11.3)
Death	3 (1.3)
Outcomes (mean mRS)	
At presentation	1.54
At follow-up	1.31

S-M: Spetzler-Martin, SD: Standard deviation

DISCUSSION

Epidemiology and natural history

CC-AVMs are relatively rare. The literature is only populated with case series and case reports; hence, the natural history of these lesions remains largely unknown. The prevalence of CC-AVMs ranges from 1.1% to 3% in population-based studies[41,42] and from 6.7% to 14.8% in hospital-based cohorts.[1,33,40] Patients with CC-AVMs present at a younger age as compared to patients with superficial AVMs. In a report by Buklina, the mean age of patients at presentation was 25 years[9] whereas it was 35 years in a series reported by Guidetti and Spallone.[18] CC-AVMs comprise 8–9% of all intracranial AVMs.[18] Since CC-AVMs are considered deep-seated lesions, the risk of hemorrhage at presentation and subsequent rehemorrhage is considered to be higher than superficial lesions, mostly by virtue of their location and deep venous drainage.[44,45,49] Our literature search yielded 37 reports describing 230 patients with CC-AVMs. Most patients harboring CC-AVMs present with ICH. Examining individual reports consisting of multiple patients revealed that 70–100% of patients present with ICH or SAH. Overall, 187 (81.3%) of 230 patients presented with hemorrhage, 10 (4%) presented with seizures without hemorrhage, and 18 (7.8%) presented with headache as the primary symptom.

Anatomical considerations

CC-AVMs are generally supplied by the branches of the anterior cerebral artery and the posterior cerebral artery (PCA) with minor contributions from the middle cerebral artery. A recent study indicated that 27 (59%) of the 46 nidi studied were supplied by both anterior and posterior circulations and 30 (65%) of 46 nidi were fed by bilateral pericallosal arteries.[37] Venous drainage generally occurs through the superior and inferior sagittal sinuses and the galenic system.

Anatomically, CC-AVMs can be divided into four main groups as described by Yasargil *et al*.[52-54] The first group includes those lesions that involve the genu and/or anterior portion of the CC. Pericallosal arteries are the typical feeders for this group of AVMs, with occasional contributions from the callosomarginal and anterior choroidal arteries. Venous drainage occurs mostly through the inferior sagittal sinus via the callosal and septal veins or the vein of Galen via internal cerebral veins. The second group of CC-AVMs consists of lesions involving the trunk of the CC; these lesions are usually fed by either unilateral or bilateral pericallosal arteries. Venous drainage occurs via the vein of Galen through the transcallosal vein; however, the inferior sagittal sinus may be involved as well. The third group consists of AVMs involving the posterior third of the body or splenium of the CC. These, too, are fed mainly by the pericallosal arteries and branches of the PCA including posterior pericallosal and posterior choroidal arteries. Venous drainage is mainly through the internal cerebral veins and vein of Galen. The fourth group consists of holocallosal AVMs, which can have variable feeders and drainage patterns. An example of a holocallosal AVM is demonstrated in Figure 1.

Picard *et al*. further divided the AVM nidi into three types such as (1) the "compact" nidus, which is well demarcated and located within the CC; (2) an "extensive" nidus involving the cingulate gyrus or septum pellucidum in addition to the CC; and (3) a "diffuse" nidus which is ill-defined and involves various cortical, subcortical, and intraventricular regions.[37]

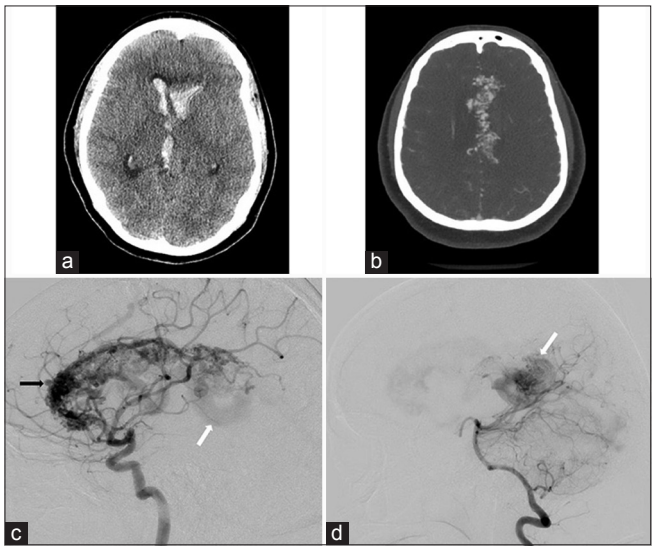

Figure 1: (a) Axial noncontrast computed tomography scan obtained on a young female patient presenting to our institution with headaches and altered mental status shows intraventricular hemorrhage. (b) Axial computed tomography angiogram performed on the same patient reveals an extensive arteriovenous malformation involving the entire corpus callosum (holocallosal). (c) Lateral projection of internal carotid artery injection reveals a large arteriovenous malformation involving the genu, rostrum, and body of the corpus callosum. Intranidal aneurysm is also appreciated (black arrow). Early venous drainage is seen via galenic system (white arrow). (d) Lateral projection of vertebral artery injection reveals filling of the splenial component of the arteriovenous malformation nidus not seen with internal carotid artery injection (white arrow)

Our analysis reveals that the most common location for CC-AVMs is the splenium. Approximately 10% of all CC-AVMs involve the white matter structure in its entirety, and these are the most difficult to treat.

Treatment

Multimodal treatment of complex AVMs is advocated. Microsurgical resections aided by endovascular embolization and stereotactic RS are the cornerstones of therapy for any AVM. Although advances in microsurgery have made complete resection of these "inoperable" lesions possible, the cure comes with a considerable risk of developing new neurological deficits. Endovascular embolization rarely achieves complete obliteration of the AVM but can significantly aid in occluding feeders that are difficult to access early in surgery. Utilization of RS for deep-seated AVMs with nidi measuring <3 cm^3 in volume has increased significantly over the last two decades with encouraging results. A more detailed discussion of these treatment modalities is as follows.

Surgical resection

Historically, surgical resection of CC-AVMs has been fraught with significant morbidity. However, with advanced microsurgical techniques and widespread availability of surgical adjuncts such as preoperative endovascular embolization and radio surgical downgrading, better results are seen in the more recent literature. One of the earliest series of surgical resection of CC-AVMs was published by Yasargil *et al*., in 1976. Eighteen patients were treated with microsurgical resection without preoperative embolization or radiotherapy.[53,54] Of the 18 patients, 15 had presented with hemorrhage and all patients belonged to a younger age group. A unilateral frontal parasagittal craniotomy and interhemispheric approach were employed in all cases for AVMs located in the anterior and middle of the CC whereas a paramedian parietal or parietooccipital craniotomy in sitting position was performed for most splenial AVMs. From a technical standpoint, the authors considered splenial AVMs to be more challenging since most of these were within the substance of the splenium and required splitting the splenium along the direction of its fibers. No operative mortality or major neurological morbidity was encountered. Hemianopsia and new onset seizures were the only postoperative complications and reported in just a few patients. Buklina undertook a complex clinical-neuropsychological study in 36 patients who underwent surgical treatment of ruptured CC-AVMs.[9] Preoperative neuropsychological testing

revealed several deficits in all patients such as memory impairment, lack of insight, impairment in optical-spatial activity, and partial "split brain" syndrome (i.e., unilateral agnosia in different modalities and impairment of transferring posture from one side to the other). In 26 of 29 cases, CC-AVM was excised via an interhemispheric approach. Any postoperative deficits were related to the specific area of the CC accessed during surgery. After surgery on the anterior portion of the CC, some patients showed worsening of frontal dysfunction, with increased disinhibition, lack of insight, and impulsivity. Split brain syndrome was more commonly observed after resection of AVM from intermediate CC whereas patients ignored the left edge of the visual field after removal of malformations from the posterior portion of the CC along with manifestation of split brain syndrome.

Bartal and Yahel argue that the incidence of paraventricular lesions, including CC-AVMs, is underestimated and needs more attention as they have a higher propensity to bleed and surgical resection could result in establishing a definitive cure.[6] They mention a case of a young girl in whom the CC-AVM initially bled and then demonstrated significant increase in size necessitating treatment. The porencephalic cyst formed by prior hemorrhage was thought to be the underlying reason for the growth of AVM. Guidetti and Spallone also noted that several recurrences of SAH in patients with these malformations was not uncommon and strongly recommended treatment.[18] Of the 4 patients in their series that were managed conservatively, 3 experienced recurrent SAH, leading to fatality of 1 patient. Conversely, outcomes of 12 patients who underwent surgical resection of the AVM were encouraging with minimal neurological or psychological deficits. Kosary *et al*. also reported excellent outcomes in 2 of 3 patients with splenial AVMs that were treated surgically.[24]

Our pooled analysis of the literature identified fifty patients who underwent surgical resection of the CC-AVM with or without prior embolization or RS, and cure was achieved in 47 (94%). Only 2 of 46 patients who underwent stand-alone surgical excision of the AVM had residual AVM on follow-up angiogram. Sixteen (32%) patients developed permanent complications after surgery, and only 1 patient died after surgical intervention.

Endovascular therapy

CC-AVMs are difficult to treat with endovascular therapy alone. The goal of embolization should be prevention of hemorrhage and a decrease in nidus size to facilitate subsequent surgical resection or RS. In 1996, Picard *et al*. described a series of AVMs involving the CC that were treated by endovascular means only.[37] They noted the difficulties in treating such lesions using endovascular therapy alone, with an overall occlusion rate of only 40%. A more recent study showed that more than half of the CC-AVMs in the series were completely obliterated using endovascular therapy alone, largely due to recent advances in the field and the advent of more sophisticated catheters and embolization material.[37,39,49] The same study also assessed the factors responsible for incomplete obliteration and found that holocallosal type, extension of the nidus to an eloquent area, nidus size >30 mm, and a Spetzler-Martin grade ≥3 were associated with failed endovascular treatment. The complication rate associated with these procedures ranges from 5% to 20%.[37,39,49] Our analysis of the literature revealed that fifty patients underwent standalone endovascular embolization of the AVM and cure was achieved in only 20 (40%), similar to the results noted by Picard *et al*. Only four patients underwent presurgical embolization of their AVMs, of which 3 (75%) were completely excised. Similarly, 26 patients underwent endovascular embolization of the AVM before radiation was delivered, but the AVM was completely occluded in only 11 (42.3%). Therefore, the authors believe that although endovascular intervention is extremely helpful in cases of large AVMs with multiple feeders or AVMs with deep or inaccessible feeders, complete obliteration is rarely achieved and microsurgery or RS must supplement embolization to achieve definitive cure.

Radiosurgery

RS is increasingly employed for treatment of small (<3 cm^3) as well as some medium- (3–6 cm) and large-size (>6 cm^3) AVMs located in deep cerebral tissue. RS is an attractive choice due to its noninvasive nature, minimal risk for acute complications, and shorter recovery time for the patients. However, cure is not immediate and can take 2–3 years for the effects of radiation to result in thrombosis of the AVM. Although smaller AVMs (<3 cm^3) are ideal candidates for RS, several reports have confirmed lower obliteration rate in larger AVMs.[22,35,36,46,51] Staged RS as well as prior embolization to aid in the reduction of radio surgical treatment volume has been utilized in the management of large AVMs not amenable to surgery.[1,14] Obliteration rate varies based on the AVM volume. Obliteration rate in AVMs with volumes between 10 and 15 cm^3 has been shown to be approximately 77%, compared to 25% for those with a volume >15 cm^3.[35] Larger AVMs also require a longer time for obliteration to occur.[55] User-friendly tools are rapidly becoming available that can guide clinicians on likelihood of AVM obliteration after RS as well as treatment-related morbidity. The RS-based grading system and the Virginia RS AVM scale were developed recently and consider AVM volume and location, patient age, and history of hemorrhage when determining a score.[38,43]

CONCLUSIONS

There is a paucity of available literature on CC-AVMs. The appropriate treatment strategy for these rare lesions remains controversial. Through this systematic review of literature and analysis of pooled data encompassing almost half a century, the authors have attempted to provide a tool that will assist healthcare providers to formulate an individualized treatment plan for these patients. Although microsurgical resection offers the most definitive treatment of these lesions, improved outcomes have been observed when microsurgery is supplemented with advances in RS and endovascular therapy. However, the decision to treat a patient with CC-AVM should weigh the natural history of these lesions against potential neurological and neuropsychological morbidity that may follow treatment.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Abla AA, Rutledge WC, Seymour ZA, Guo D, Kim H, Gupta N, *et al.* A treatment paradigm for high-grade brain arteriovenous malformations: Volume-staged radiosurgical downgrading followed by microsurgical resection. J Neurosurg 2015;122:419-32.
2. Akimoto H, Komatsu K, Kubota Y. Symptomatic *de novo* arteriovenous malformation appearing 17 years after the resection of two other arteriovenous malformations in childhood: Case report. Neurosurgery 2003;52:228-31.
3. Al-Shahi R, Fang JS, Lewis SC, Warlow CP. Prevalence of adults with brain arteriovenous malformations: A community based study in Scotland using capture-recapture analysis. J Neurol Neurosurg Psychiatry 2002;73:547-51.
4. Andreussi L, Borella F, Cama A, Marino C. Microsurgical excision of an arteriovenous malformation of the anterior corpus callosum. Surg Neurol 1978;10:97-9.
5. Baiz TC, Jakoby RK. Surgical treatment of an arteriovenous malformation associated with agenesis of the corpus callosum. J Neurosurg 1964;21:306-8.
6. Bartal AD, Yahel MA. Total excision of an arteriovenous malformation of the corpus callosum. Case report. J Neurosurg 1970;33:95-9.
7. Bassett RC. Surgical experiences with arteriovenous anomalies of the brain. J Neurosurg 1951;8:59-74.
8. Bendavid OJ, Khoshyomn S, Wilson JT. Arteriovenous malformation of the genu of the corpus callosum. Pediatr Neurosurg 2004;40:49-50.
9. Buklina SB. The corpus callosum, interhemisphere interactions, and the function of the right hemisphere of the brain. Neurosci Behav Physiol 2005;35:473-80.
10. Buklina SB. The unilateral spatial neglect phenomenon in patients with arteriovenous malformations of deep brain structures. Neurosci Behav Physiol 2002;32:555-60.
11. Castro-Caldas A, Poppe P, Antunes JL, Campos J. Partial section of the corpus callosum: Focal signs and their recovery. Neurosurgery 1989;25:442-7.
12. Cone JD, Maravilla KR, Cooper PR, Diehl JT, Clark WK. Computed tomography findings in ruptured arteriovenous malformations of the corpus callosum. J Comput Assist Tomogr 1979;3:478-82.
13. Da Pian R, Pasqualin A, Scienza R, Vivenza C. Microsurgical treatment of ten arteriovenous malformations in critical areas of the cerebrum. J Microsurg 1980;1:305-20.
14. Dawson RC 3rd, Tarr RW, Hecht ST, Jungreis CA, Lunsford LD, Coffey R, *et al.* Treatment of arteriovenous malformations of the brain with combined embolization and stereotactic radiosurgery: Results after 1 and 2 years. AJNR Am J Neuroradiol 1990;11:857-64.
15. Dikel TN, Fennell EB, Nadeau SE, Quisling RG, Mickle JP, Friedman WA. A neuropsychological outcome study of a child's left pericallosal arteriovenous malformation with occult fornix lesion. Neurocase 2001;7:503-13.
16. Ganapathy K, Shankarnarayanan V, Saji, Padmanabhan TK, Halbe S. Obliteration of giant corpus callosum AVM with linac based stereotactic radiosurgery. J Clin Neurosci 2003;10:272-6.
17. Garza-Mercado R, Cavazos E, Tamez-Montes D. Cerebral arteriovenous malformations in children and adolescents. Surg Neurol 1987;27:131-40.
18. Guidetti B, Spallone A. The management of arteriovenous malformations of the corpus callosum. Neurol Res 1982;4:253-82.
19. Herzig R, Burval S, Vladyka V, Janouskova L, Krivanek P, Krupka B, *et al.* Familial occurrence of cerebral arteriovenous malformation in sisters: Case report and review of the literature. Eur J Neurol 2000;7:95-100.
20. Houtteville JP, el Omeiri S, Derlon JM, Toumi K, Benazza A. Arteriovenous aneurysm of the corpus callosum with normal arteriography 9 years earlier. Neurochirurgie 1989;35:15-22.
21. Juhász J. Surgical treatment of arteriovenous angiomas localised in the corpus callosum, basal ganglia and near the brain stem. Acta Neurochir (Wien) 1978;40:83-101.
22. Kemeny AA, Dias PS, Forster DM. Results of stereotactic radiosurgery of arteriovenous malformations: An analysis of 52 cases. J Neurol Neurosurg Psychiatry 1989;52:554-8.
23. Kohmura E, Taki T, Tanioka T. Multiple intracerebral arteriovenous malformations in deep structure – Case report. Neurol Med Chir (Tokyo) 1990;30:624-7.
24. Kosary IZ, Braham J, Shacked I, Kronenberg Y. Vascular malformation of the posterior corpus callosum: Surgical treatment. Surg Neurol 1978;10:345-7.
25. Koyanagi I, Abe H, Nakagawa Y, Miyamachi K, Sasaki H, Miyasaka K, *et al.* Nidus embolization of cerebral arteriovenous malformation fed mainly by a pericallosal artery prior to surgical excision. Case report. No Shinkei Geka 1985;13:1019-24.
26. Kunc Z. Arteriovenous malformations in the corpus callosum. Cesk Neurol Neurochir 1974;37:253-7.
27. Lobato RD, Gómez PA, Lagares A, Campollo J, González P, Boto GR, *et al.* Parasplenial arteriovenous malformations. Report of 15 surgically treated cases. Neurocirugia (Astur) 2002;13:15-21.
28. Machado de Almeida G, Shibata MK, Nakagawa EJ. Contralateral parafalcine approach for parasagittal and callosal arteriovenous malformations. Neurosurgery 1984;14:744-6.
29. Maruyama K, Shin M, Tago M, Kurita H, Kawamoto S, Morita A, *et al.* Gamma knife surgery for arteriovenous malformations involving the corpus callosum. J Neurosurg 2005;102 Suppl:S49-52.
30. McDonald CR, Crosson B, Valenstein E, Bowers D. Verbal encoding deficits in a patient with a left retrosplenial lesion. Neurocase 2001;7:407-17.
31. Milhorat TH. Excision of a cirsoid arteriovenous malformation of the corpus callosum in a 16-year-old boy. Case report. J Neurosurg 1970;33:339-44.
32. Mohanty CB, Devi BI, Somanna S, Bhat DI, Dawn R. Corpus callosum arteriovenous malformation with persistent trigeminal artery. Br J Neurosurg 2011;25:736-40.
33. Moher D, Liberati A, Tetzlaff J, Altman DG; PRISMA Group. Preferred reporting items for systematic reviews and meta-analyses: The PRISMA statement. Int J Surg 2010;8:336-41.
34. Orozco LD, Luzardo GD, Buciuc RF. Transarterial balloon assisted onyx embolization of pericallosal arteriovenous malformations. J Neurointerv Surg 2013;5:e18.
35. Pan DH, Guo WY, Chung WY, Shiau CY, Chang YC, Wang LW. Gamma knife radiosurgery as a single treatment modality for large cerebral arteriovenous malformations. J Neurosurg 2000;93 Suppl 3:113-9.
36. Pendl G, Unger F, Papaefthymiou G, Eustacchio S. Staged radiosurgical treatment for large benign cerebral lesions. J Neurosurg 2000;93 Suppl 3:107-12.
37. Picard L, Miyachi S, Braun M, Bracard S, Per A, Marchal JC. Arteriovenous malformations of the corpus callosum – Radioanatomic study and effectiveness of intranidus embolization. Neurol Med Chir (Tokyo) 1996;36:851-9.
38. Pollock BE. Development and testing of a radiosurgery-based arteriovenous malformation grading system. Prog Neurol Surg 2013;27:58-66.
39. Robert T, Blanc R, Ciccio G, Gilboa B, Fahed R, Redjem H, *et al.* Angiographic factors influencing the success of endovascular treatment of arteriovenous malformations involving the corpus callosum. J Neurointerv Surg 2015;7:715-20.
40. Sell SC. The corpus callosum, its role in memory: A presentation of a patient with an arteriovenous malformation of the corpus callosum. J Neurosurg Nurs 1977;9:141-3.

41. Shi YQ, Chen XC. Arteriovenous malformation of corpus callosum. Results of surgical treatment in 5 cases. Chin Med J (Engl) 1987;100:87-91.
42. Shimizu S, Irikura K, Miyasaka Y, Mochizuki T, Kurata A, Kan S, *et al.* Rupture of pial arteriovenous malformation associated with early thrombosis of the draining system following stereotactic radiosurgery – Case report. Neurol Med Chir (Tokyo) 2001;41:599-602.
43. Starke RM, Yen CP, Ding D, Sheehan JP. A practical grading scale for predicting outcome after radiosurgery for arteriovenous malformations: Analysis of 1012 treated patients. J Neurosurg 2013;119:981-7.
44. Stefani MA, Porter PJ, terBrugge KG, Montanera W, Willinsky RA, Wallace MC. Angioarchitectural factors present in brain arteriovenous malformations associated with hemorrhagic presentation. Stroke 2002;33:920-4.
45. Stefani MA, Porter PJ, terBrugge KG, Montanera W, Willinsky RA, Wallace MC. Large and deep brain arteriovenous malformations are associated with risk of future hemorrhage. Stroke 2002;33:1220-4.
46. Steiner L, Leksell L, Greitz T, Forster DM, Backlund EO. Stereotaxic radiosurgery for cerebral arteriovenous malformations. Report of a case. Acta Chir Scand 1972;138:459-64.
47. Uchino A, Matsunaga M, Ohno M. Arteriovenous malformation of the corpus callosum associated with persistent primitive trigeminal artery – Case report. Neurol Med Chir (Tokyo) 1989;29:429-32.
48. Valenstein E, Bowers D, Verfaellie M, Heilman KM, Day A, Watson RT. Retrosplenial amnesia. Brain 1987;110(Pt 6):1631-46.
49. Viñuela F, Dion JE, Duckwiler G, Martin NA, Lylyk P, Fox A, *et al.* Combined endovascular embolization and surgery in the management of cerebral arteriovenous malformations: Experience with 101 cases. J Neurosurg 1991;75:856-64.
50. Wang YC, Lee SD, Chen NF, Shen CC. Cerebral intraventricular hemorrhage caused by a large cerebral arteriovenous malformation at 31 years after diagnosis. Zhonghua Yi Xue Za Zhi (Taipei) 2001;64:121-8.
51. Yamamoto M, Hara M, Ide M, Ono Y, Jimbo M, Saito I. Radiation-related adverse effects observed on neuro-imaging several years after radiosurgery for cerebral arteriovenous malformations. Surg Neurol 1998;49:385-97.
52. Yasargil MG. Microneurosurgery: AVM of the Brain, History, Embryology, Pathological Considerations, Hemodynamics, Diagnostic Studies, Microsurgical Anatomy. 1st ed. New York: Georg Thieme Verlag, Thieme Medical Publishers, Inc.; 1987.
53. Yasargil MG, Jain KK, Antic J, Laciga R. Arteriovenous malformations of the splenium of the corpus callosum: Microsurgical treatment. Surg Neurol 1976;5:5-14.
54. Yasargil MG, Jain KK, Antic J, Laciga R, Kletter G. Arteriovenous malformations of the anterior and the middle portions of the corpus callosum: Microsurgical treatment. Surg Neurol 1976;5:67-80.
55. Zabel-du Bois A, Milker-Zabel S, Huber P, Schlegel W, Debus J. Stereotactic linac-based radiosurgery in the treatment of cerebral arteriovenous malformations located deep, involving corpus callosum, motor cortex, or brainstem. Int J Radiat Oncol Biol Phys 2006;64:1044-8.