A study of pediatric cerebral arteriovenous malformations: clinical presentation, radiological features, and long-term functional and educational outcomes with predictors of sustained neurological deficits

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OBJECTIVE Large experiences with the treatment of pediatric arteriovenous malformations (AVMs) remain relatively rare, with limited data on presentation, treatment, and long-term functional outcomes. Because of the expected long life-span of children, caregivers are especially interested in outcome measures that assess quality of life. The authors' intention was to describe the long-term functional outcomes of pediatric patients who undergo AVM surgery and to identify predictors of sustained neurological deficits.

METHODS The authors analyzed a 21-year retrospective cohort of pediatric patients with intracranial AVMs treated with microsurgery at two institutions. The primary outcome was a persistent neurological deficit at last follow-up. Secondary outcome measures included modified Rankin Scale (mRS) score and independent living.

RESULTS Overall, 97 patients (mean age 11.1 ± 4.5 years; 56% female) were treated surgically for intracranial AVMs (mean follow-up 77.5 months). Sixty-four patients (66%) presented with hemorrhage, and 45 patients (46%) had neurological deficits at presentation. Radiologically, 39% of lesions were Spetzler-Martin grade II. Thirty-seven patients (38%) with persistent neurological deficits at last follow-up were compared with those without deficits; there were no differences in patient age, presenting Glasgow Coma Scale score, AVM size, surgical blood loss, or duration of follow-up. Multivariate analysis demonstrated that a focal neurological deficit on presentation, AVM size > 3 cm, and lesions in eloquent cortex were independent predictors of persistent neurological deficits at long-term follow-up. Overall, 92% of the children had an mRS score ≤ 2 on long-term follow-up.

CONCLUSIONS Pediatric patients with AVMs treated with microsurgical resection have good functional and radiological outcomes. There is a high rate (38%) of persistent neurological deficits, which were independently predicted by preoperative deficits, AVMs > 3 cm, and lesions located in eloquent cortex. This information can be useful in counseling families on the likelihood of long-term neurological deficits after cerebral AVM surgery.

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KEYWORDS arteriovenous malformations; deficit; pediatric; Spetzler-Martin grading system; vascular disorders

ABBREVIATIONS aOR = adjusted odds ratio; AVM = arteriovenous malformation; FND = focal neurological deficit; GCS = Glasgow Coma Scale; mRS = modified Rankin Scale.

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INTRACRANIAL arteriovenous malformations (AVMs) in children account for up to 55% of pediatric hemorrhagic strokes.^{2,5,6,11,13,24} Large experiences with modern treatment modalities coupled with long-term follow-up remain limited. The expected long lifespans of children mean that caregivers are especially interested in outcome measures that directly affect quality of life, including the likelihood of persistent focal neurological deficits (FNDs), the probability of clinical improvement (or decline) over time, and quality-of-life outcomes such as ability to achieve independent living and educational status.

Using a large cohort from two institutions, we examined the clinical presentation, radiological features, and treatment of pediatric patients with intracranial AVMs managed by microsurgical resection. We focused on long-term outcomes with the specific goal of identifying clinical and radiological AVM factors that predict persistent FNDs. We hypothesized that certain clinical and radiological factors might predict long-term functional and neurological outcomes.

Methods

Study Design

This study was a retrospective review of prospectively maintained surgical databases at two pediatric hospitals: Boston Children's Hospital (MA) and Primary Children's Hospital (Salt Lake City, UT). Separate IRB approval was obtained from our institutions with a waiver of informed consent. The article was prepared with the STROBE guidelines.²⁰

Inclusion/Exclusion Criteria

We identified consecutive patients ≤ 18 years of age who were diagnosed with an intracranial AVM and who were treated with resection from May 1996 through February 2017. We reviewed clinical data and radiological records available from the two institutions for each patient. Patients with arteriovenous fistulas, vein of Galen malformations, and spinal vascular lesions were excluded. Patients with < 12 months of follow-up were excluded from the analysis.

Data Collection

Clinical variables included age at surgery, presence of associated hereditary syndromes, other existing medical diagnoses, presenting symptoms and Glasgow Coma Scale (GCS) score, modified Rankin Scale (mRS) score,²⁶ any FNDs on presentation, headache, seizures, or intracranial hemorrhage.

Radiological data included review for hemorrhage at presentation, hydrocephalus requiring external ventricular drainage, lesion size > 3 cm in diameter, lesion location, and key angioarchitectural characteristics (location, size, venous drainage, associated nidal aneurysms, and feeding artery aneurysms).

Treatment data included use of preoperative embolization, iatrogenic sequelae, and timing of surgery.

Radiological outcomes included AVM obliteration after surgery and the development of postoperative residual or recurrent lesions. At each center, the follow-up protocol

involves obtaining a brain MR image at 6 months, angiogram at 1 year, and yearly brain MR images up to 5 years after surgery.

Clinical outcomes included length of postoperative follow-up, changes in mRS score, presence of new or persisting neurological deficits, persistence of headaches, interval development of new seizures, or repeat hemorrhage. Highest level of education and status of independent living were also assessed via delayed telephone interview by trained research assistants.

Statistical Analysis

We compared children with and without persistent FNDs. Data were summarized using means and standard deviations for continuous variables (Mann-Whitney U-test) and counts and frequencies for categorical variables (Fisher exact test). Univariate statistical analysis was performed to identify the unadjusted association of demographic variables, clinical signs, and radiological characteristics with persistent FND.

Multivariate logistic regression analysis was performed by including variables satisfying statistical entry criteria ($p < 0.2$) on univariate analysis, as well as clinical factors of interest regardless of statistical association in our univariate analysis. Calibration was examined using the Hosmer-Lemeshow goodness-of-fit test. Statistical significance was established using a value of $p < 0.05$. Data were analyzed using SAS software.

Results

Clinical Presentation

During the 21-year study period, 150 pediatric patients were treated surgically for a confirmed diagnosis of cerebral AVM at these 2 children's hospitals. A total of 97 patients (mean age 11.1 ± 4.5 years, 56% female) satisfied the inclusion criteria and had a minimum of 12 months of follow-up—78% from one institution and 22% from another. The median follow-up duration for the entire cohort was 65 months (range 12–292 months). For children without an FND, the median follow-up duration was 64 months (range 12–201 months), and for children with an FND, the median follow-up duration was 70 months (range 12–292 months). Fifty-three patients were excluded because of lack of clinical and radiological follow-up. Six patients (6.2%) had a diagnosis of hereditary hemorrhagic telangiectasia, and 9 (9.3%) had other known preexisting medical conditions. Sixty-four patients (66.0%) presented with hemorrhage, 64 (66.0%) with headache, and 15 (15.5%) with seizures. Approximately half of the children ($n = 45$, 46.4%) had new neurological deficits at presentation, and the mRS score was > 2 in 34 patients (35.1%) (Table 1). Other clinical presentation data are provided in Tables 1 and 2.

Radiological Presentation

The AVM location was most commonly supratentorial ($n = 86$, 88.7%) (Table 3). Approximately two-thirds ($n = 64$, 66.0%) of the patients had intracranial hemorrhage on initial scan. Using the Spetzler-Martin grading scale,²⁵ 29 (29.9%) of the AVMs were grade I, 36 (37.1%) were grade II, 20 (20.6%) were grade III, 7 (7.2%) were grade IV, and

TABLE 1. Summary of categorical variables using the Fisher exact test

Variable	No Persistent FND (n = 60)	Persistent FND (n = 37)	p Value
Clinical/presentation characteristics			
Female	36/60 (60)	18/37 (49)	0.3
HHT	5/60 (8)	1/37 (2.7)	0.40
Symptomatic (headache, seizure, hemorrhage, etc.)	51/60 (80)	34/37 (92)	0.36
Other medical disease	6/60 (10)	3/37 (8.1)	1.00
mRS score >2 on presentation	17/59 (29)	17/37 (46)	0.124
FND on presentation	19/60 (32)	26/37 (70)	<0.001
Visual field cut	7/60 (12)	8/37 (22)	0.25
Hemiparesis/hemiplegia	12/60 (20)	20/37 (54)	<0.001
Cranial nerve deficit	4/60 (6.6)	5/37 (14)	0.29
Hemorrhage	40/60 (67)	24/37 (65)	1.00
Headache	39/60 (65)	21/37 (57)	0.52
GCS score ≤8	9/59 (15)	9/37 (24)	0.29
Seizures	8/60 (13)	7/37 (19)	0.57
Embolization	18/60 (30)	17/37 (46)	0.13
Bleed with embolization	1/60 (1.6)	2/37 (5.4)	0.56
Urgent op (acutely following rupture)	24/60 (40)	11/37 (30)	0.386
Radiological factors			
Ventricular enlargement/hydrocephalus requiring urgent EVD	9/60 (15)	10/37 (27)	0.19
Mass effect	15/59 (25)	13/36 (36)	0.35
Size >3 cm in diameter	10/59 (17)	14/35 (40)	0.016
Lobar location	31/59 (53)	23/35 (66)	0.28
Cerebellar	10/60 (17)	2/37 (5.4)	0.12
Eloquent cortex	19/60 (32)	25/37 (68)	<0.001
Deep drainage	17/60 (28)	16/36 (44)	0.12
Nidal aneurysm	3/60 (5)	4/37 (11)	0.42
Angiographically confirmed obliteration	50/60 (83)	29/37 (78)	0.59
Interval recurrence	9/59 (15)	7/37 (19)	0.78
Long-term clinical outcomes			
Survival at 5 yrs*	35/36 (97)	23/23 (100)	1.00
Living independently (ADLs)	55/59 (93)	28/37 (76)	0.028
Visual field cut	2/59 (3.3)	15/37 (41)	<0.001
Hemiparesis/hemiplegia	1/59 (1.6)	21/37 (57)	<0.001
Cranial nerve deficit	0/59 (0)	7/37 (19)	0.001
Headaches	8/59 (14)	6/37 (16)	0.771
New seizures	9/59 (15)	8/37 (22)	0.427
Rebleed	1/60 (1.6)	0/37 (0)	1.00

ADL = activities of daily living; EVD = external ventricular drainage; HHT = hereditary hemorrhagic telangiectasia.

Boldface type indicates statistical significance.

* Of patients with ≥ 5 years of follow-up (total n = 59).

1 (1.0%) was grade V, with significant differences in the distribution of grades between the two cohorts ($p = 0.012$) (Table 4). Most AVMs (75.3%) were small (≤ 3 cm), with a mean size of 2.4 ± 1.22 cm (in greatest axis). Thirty-three (34.0%) AVMs had deep venous drainage, and 7 (7.2%) patients had an associated aneurysm on an associated feeding artery or at an intranidal location; 44 (45.4%) lesions were located in eloquent cortex (Table 1).

Treatment Data

All patients underwent AVM resection, with 35 (36.1%) requiring an emergent operation (including clot evacuation for increased intracranial pressure) at presentation. Nineteen patients (19.6%) required placement of an external ventricular drain for hydrocephalus. Thirty-five patients (36.1%) underwent preoperative embolization. The mean estimated blood loss for surgery was 252 ml. Complica-

TABLE 2. Summary of continuous variables using the Mann-Whitney U-test

Variable	No Persistent FND			With Persistent FND			p Value
	No.	Mean $\pm$ SD	25/50/75th Percentile	No.	Mean $\pm$ SD	25/50/75th Percentile	
Age at op (mean in yrs)	60	11.1 $\pm$ 4.35	8.23/11.5/14.5	37	11.4 $\pm$ 4.53	9.08/11/13.9	0.77
Presenting GCS score	59	13.1 $\pm$ 3.6	14/15/15	37	12.1 $\pm$ 4.2	8.5/15/15	0.16
Lesion size (cm)	57	2.3 $\pm$ 1.08	1.6/2/2.55	33	2.81 $\pm$ 1.5	1.85/2.5/3.45	0.16
Time to FU (mos)	60	76.5 $\pm$ 51.6	30.5/64/112	37	80.3 $\pm$ 63	36.5/70/101	0.98
Estimated blood loss (ml)	50	265 $\pm$ 304	100/150/363	34	242 $\pm$ 256	100/150/263	0.88
Mos until FU imaging	59	51.8 $\pm$ 45.9	16/34/77	37	56.7 $\pm$ 44.4	25.5/48/72.5	0.31

FU = follow-up.

tions occurred in 3 (8.6%) of 35 embolization procedures (all hemorrhages). There were no intraoperative deaths.

Radiological and Clinical Outcome Data

Seventy-nine patients (81.4%) had angiographically confirmed obliteration of the AVM after surgery (within the operative hospitalization). There was an association between having recurrent/residual lesions identified on delayed angiography and not having undergone postoperative angiography during the intraoperative or immediate postoperative period ($p < 0.001$). Sixteen patients had an interval recurrence (Table 1), only 4 of whom had confirmed obliteration on immediate postoperative angiography. Twelve (75%) of the 16 patients who had recurrent lesions initially presented with acute AVM hemorrhage. Nine of 16 patients were taken for urgent surgery (< 24 hours). One patient experienced a repeat hemorrhage from a recurrent AVM 15 years after resection.

Clinically, of the 97 patients with long-term follow-up (mean 77.5 months), 37 (38.1%) had a persistent FND at last follow-up. Five patients (17.2%) with Spetzler-Martin grade I lesions, 13 (36.1%) with grade II lesions, 10 (50%) with grade III, 5 (71.4%) with grade IV lesions, and the only patient (100%) with a Spetzler-Martin grade V lesion had a persistent FND on follow-up. Specific deficits are quantified in Table 1.

Fifteen patients had visual field deficits preoperatively, and 17 had visual deficits postoperatively. There were 8 persistent visual field deficits (6 occipital, 1 temporal, and 1 parietal), 7 resolved visual field cuts, and 9 new visual field cuts (4 occipital, 4 temporal, and 1 thalamic). Hemiparesis/hemiplegia persisted in 16 patients and resolved on long-term follow-up in 16 patients. Six patients developed new hemiparesis/hemiplegia (2 parietal and 1 each

occipital, temporal, cerebellar, and thalamic). There were 9 cranial nerve deficits preoperatively: 7 resolved and 2 persisted. There were 5 new cranial nerve deficits postoperatively that persisted until last follow-up (Table 5).

Overall, 58 (98.3%) of 59 patients were alive at ≥ 5 -year follow-up, with 1 death from a residual AVM hemorrhage. Eighty-three patients (85.5%) were living independently at last follow-up. Forty-one patients (mean age at surgery 12 ± 4 years; mean follow-up duration 75.8 ± 51 months) were available to complete telephone interviews regarding education status (Table 6): 7.3% completed or were enrolled currently in grade school, 51.2% completed high school, 29.2% completed college, 7.3% completed professional school, and 4.9% were not in school because of poor grades. Overall, 92% of children did well, with an mRS score ≤ 2 on long-term follow-up. The delta mRS score (change from initial presentation to last follow-up) was calculated for each available patient (Fig. 1). Approximately 47% of the children had an improvement in their mRS score over time, 36% had a stable mRS score, and only 11% had a worsened mRS score. Eighty-six (86.5%) of 93 patients (no deficit [93%] vs deficit [76%], $p = 0.03$) were living independently at their last follow-up.

Identifying Risk Factors Predictive of Sustained Deficits

When comparing patients with and without persistent FNDs at last follow-up, there was no difference in mean age, presenting GCS score, AVM size, operative blood loss, or length of follow-up between the two cohorts (Table 1). Univariate analysis demonstrated that FNDs on presentation (32% vs 70%, $p < 0.001$), specifically hemiparesis or hemiplegia (20% vs 54%, $p < 0.001$), size > 3 cm in diam-

TABLE 3. Location of AVMs in 97 patients

Location	No. of Patients
Temporal	18
Frontal	22
Parietal	18
Occipital	19
Cerebellar	12
Other (thalamic, brainstem)	8

TABLE 4. Spetzler-Martin graded of 93 surgically treated AVMs with data available

Spetzler-Martin Grade	No Persistent FND (n = 59)	Persistent FND (n = 34)
I	24 (41%)	5 (15%)
II	23 (39%)	13 (38%)
III	10 (17%)	10 (29%)
IV	2 (3.4%)	5 (15%)
V	0 (0%)	1 (2.9%)

Values are presented as the number (%) of AVMs.

TABLE 5. Type of neurological deficits on initial presentation and postoperatively in 97 patients

Deficit	Initial Presentation	Postoperatively
FND	45 (46%)	37 (38%)
Visual field deficit	15 (15%)	17 (18%)
Hemiparesis/hemiplegia	32 (33%)	22 (23%)
Cranial nerve deficit	9 (9%)	7 (7%)

Values are presented as the number (%) of patients. Some patients had more than one deficit so numbers do not total 100%.

eter (17% vs 38%, $p = 0.016$), and lesions in eloquent cortex (32% vs 68%, $p < 0.001$) had an unadjusted association with persistent FNDs on long-term follow-up (Table 2).

The multivariate regression analysis demonstrated that an FND on presentation (adjusted OR [aOR] 7.43 [95% CI 2.00–27.5]), AVM size > 3 cm in diameter (aOR 2.97 [95% CI 0.93–9.586], and lesions in eloquent cortex (aOR 2.67 [95% CI 0.964–7.44]) were independent predictors of persistent FND at long-term follow-up (Table 7). The Hosmer-Lemeshow statistic was not significant, consistent with adequate model calibration.

Discussion

We present a large cohort of intracranial AVMs surgically treated at two centers in children with long-term follow-up. Previous reports of treatment outcomes have been limited by smaller cohort size.^{4,5,10,11,15,19,22} Our data clarify the clinical and radiological presentation of this population and provide perioperative and long-term outcomes. We identified 3 independent risk factors for persistent FND in children who undergo surgical treatment for cerebral AVMs. An FND on presentation, AVM size > 3 cm, and cerebral AVMs located in eloquent cortex were independently associated with persistent FNDs on long-term follow-up. These data will help to inform the treatment of children with AVMs when considering resection.

Importantly, this study differentiates itself from others by providing a novel approach for examining and predicting long-term functional outcomes for children and by providing potential information when counseling children and families on long-term neurological deficits in the setting of cerebral AVM surgery.

Novelty of the Study and Follow-Up

In addition to increasing the overall quantity of data (in terms of both patient number and follow-up length), our results reveal 4 important findings relevant to the outcomes after surgical management of pediatric AVMs.

First, our study improves our understanding of the evolving role of perioperative embolization as an adjunct in treating cerebral AVMs before surgery. Although there was an 8.5% rate of hemorrhagic complications during embolization, they did not correlate with focal deficits on follow-up, indicating that embolization is a safe treatment adjunct in the long term. Second, this multicenter cohort highlights the overall good long-term outcomes of these patients, with stable or improving mRS scores over time,

TABLE 6. Education level attained by children after surgical treatment for AVM (n = 41 respondents)

Level of Education*	Frequency	% of Those Responding
Grade school	3	7.3
High school	21	51.2
College	12	29.2
Professional school	3	7.3
Not in school due to poor grades	2	4.9

* Varying ages of respondents.

high rates of independent living, and advanced educational success. Third, this work reinforces the importance of high-quality intra- or perioperative angiography to confirm AVM obliteration. In 16 patients with recurrence, only 4 had confirmed obliteration on immediate postoperative angiography; the rest did not undergo angiography. Thus, the recurrences may have resulted from failure to recognize incomplete resection or residual lesion. Fourth, our results offer insights into the risks of emergent operation. Twelve (75%) of 16 recurrences initially presented with acute hemorrhage, and 9 had urgent surgery (< 24 hours); thus, there is a possibility that acute hemorrhage or cerebral edema skewed the recognition of the entire vascular lesion, thus contributing to the interval recurrence rate.

Equally important to understanding the patterns and risks for persistent FNDs is the need for dedicated long-term clinical and radiological follow-up for these children. There is a risk of recurrence, clinical deterioration, and rehemorrhage. In treating pediatric AVMs, counseling patients and families about treatment options, procedural morbidity, and obliteration rates is critical.⁹ Our data help to clarify factors that influence the risk of long-term neurological disability, affording physicians specific tools—perioperative angiography, for example—and identifiable risk factors (such as emergent operation vs elective resection) that can now guide treatment and follow-up with greater accuracy.

FIG. 1. Bar graph demonstrating the change in mRS scores from initial presentation to last follow-up.

TABLE 7. Multivariate analysis for independent risk factors associated with persistent neurological deficits after AVM resection

Variable	aOR	95% CI	p Value
FND on presentation	7.43	2.00–27.5	0.003
Size >3 cm in diameter	2.97	0.93–9.586	0.06
Eloquent cortex	2.67	0.964–7.44	0.05
mRS score >2 on presentation	0.311	0.311–3.69	0.91
Hemorrhage on presentation	0.57	0.13–2.50	0.45
Seizure on presentation	1.58	0.38–6.53	0.53

Boldface type indicates statistically significant values ($p < 0.05$).

ARUBA Study and Rebleeding Rates

Our investigation has additional utility when viewed within the context of the recent ARUBA (A Randomized Trial of Unruptured Brain AVMs) study.¹⁸ Our current study challenges the ARUBA data—at least for children—by demonstrating the predictors of neurological morbidity and showing that children do well across several outcome metrics (mRS score, independence, and education) after microsurgical treatment. The additional lifelong cumulative risk of rupture and the potential for morbidity from AVM hemorrhage are important factors that justify treatment in appropriately selected patients. The Scottish Audit of Intracranial Vascular Malformations supported the findings of the ARUBA study but also judged that cumulative rates of death or handicap with follow-up > 12 years may favor treatment.¹ Whereas the ARUBA and Scottish Audit of Intracranial Vascular Malformations studies were focused on adult unruptured AVMs, the natural history and longevity in children demonstrate a need for different treatment thresholds in pediatric patients.

Interestingly, hemorrhage was not significantly associated with the primary outcome. Although some previous reports have demonstrated lower neurological complication rates,^{7,17} the overall neurological and long-term clinical outcomes in this cohort study are quite good. No comparative data exist for larger series of patients treated with embolization only or radiosurgery. Delayed cognitive sequelae of radiation therapy are of interest, especially in the pediatric population.^{8,12,21,23} Our data also support the routine use of immediate postoperative angiography to ensure AVM obliteration, given the association with residual lesions and not obtaining an angiogram in the immediate postoperative period ($p < 0.001$). Although Blauwblomme et al.³ demonstrated that associated aneurysms and deep venous drainage as AVM characteristics were independent risk factors for rebleeding in pediatric AVMs, we did not find these factors to be associated with persistent deficit.

Long-Term Functional Outcomes

Long-term functional outcomes are of particular interest when treating pediatric patients. We attempted to augment standardized outcome metrics (such as the mRS score) with more intuitive data that can help inform patients and families with a more complete picture when predicting outcomes. An overwhelmingly majority of patients

(92%) had an mRS score of 0–2 on long-term follow-up, indicating a durable result. Worsening mRS score, which was seen in a small subset of patients in this series, may occur in the setting of recurrent seizure activity secondary to AVM rupture, hemosiderin, persistent headaches, or cognitive dysfunction, which could be a result of AVM rupture at an early age with long-term sequelae.

More relatable data included overall survival, independent living, and education status. For patients with ≥ 5 years of follow-up, only one death (1.7%) occurred, related to a residual AVM. Overall, 86% (83 of 97) of the patients were living independently at long-term follow-up. When discussing outcomes, the long-term educational potential for children who have undergone craniotomy for pediatric AVMs is of significant interest. Overall, 29% of respondents completed a college-level education, with 7.3% achieving professional school completion. We believe outcome measures such as educational achievement, neuropsychological testing, and intelligence testing should be more commonplace in pediatric studies because of the longevity and growth occurring in this population.

Recurrent AVMs in Pediatric Patients

In this series, 16 patients had an interval recurrence identified on follow-up imaging. Not having an immediate angiogram, acute hemorrhage, and the need for urgent surgery likely contributed to the recurrence rate in this series. In fact, 12 of 16 patients with recurrent lesions did not undergo immediate postoperative angiography; thus, these recurrences may in fact represent residual lesions rather than recurrent ones. The rates of recurrence tend to be higher in children (13%–14%) than in adults even after complete obliteration,^{15,16} because AVM formation is an “active process” and immature blood vessels in the resection cavity may predispose children to recurrent AVM formation.¹⁴ Thus, close short- and long-term radiological surveillance is necessary for children with AVMs, despite a “clean” immediate postoperative angiogram, even though a number of patients did not have an immediate completion angiogram. Each center currently follows a protocol that calls for surveillance for 5 years; given that one patient in this investigation experienced a recurrence at > 5 years, yearly surveillance for a longer period may be necessary.

Cause of Neurological Deficits

Overall, 45 patients (46%) presented with an FND (Table 7). We surmise that a majority of the neurological deficits come from the initial ictus of hemorrhage and are spontaneous in origin. The information in Table 7 helps in the understanding of neurological deficits and their propensity to improve over time. It is difficult to make assumptions about the prevalence and persistence of visual field cuts as some may have resolved and been a result of surgical approach and resection (majority occipital lobe lesions; 75%). Thus, the results of persistent long-term deficits should be taken in this light. Despite the presence of these deficits, however, the mRS score and independence with respect to activities of daily living seem to be unaffected, allowing a potentially excellent quality of life.

Limitations

We conducted a retrospective multicenter study with inherent recall bias. The most notable limitation is the elimination of 53 patients based on limited follow-up information. Although both centers have recommended follow-up paradigms, the tertiary nature of each of the centers drawing patients from long distances means that follow-up information can be difficult to obtain in a reliable fashion. We chose to include patients with only 12 months of follow-up as the most responsible manner to undertake the study given the hypothesis and objectives. Current treatment of AVMs includes surgery, stereotactic radiosurgery, endovascular embolization, or a combination of these. In this study, we only included patients treated initially with surgery with/without embolization as an adjunct. Information collected as a proxy for cognitive outcome included a telephone questionnaire for a fraction of the study cohort and thus is subject to bias. In addition, the varying ages of respondents and ages of initial surgical intervention make direct comparison of these metrics difficult.

There are inherent shortcomings when assessing independent functionality in children, specifically with regard to the mRS score. Although educational level can be used as a measure for cognitive function, the variability in follow-up and age ranges among the respondents limit the conclusions drawn from the data presented. Despite these limitations, we believe this study provides valuable information on the long-term clinical outcomes and predicting persistent FNDs in children undergoing surgery for cerebral AVMs and can be useful in counseling of children and families on the long-term neurological deficits in the setting of cerebral AVM surgery.

Conclusions

Pediatric AVMs treated with microsurgical resection have good functional and radiological outcomes, particularly when perioperative angiograms are used to confirm obliteration. We identified 3 presentation characteristics that were independently associated with persistent FNDs on long-term assessment after surgical treatment for cerebral AVMs in children: FND on initial presentation, AVM size > 3 cm, and AVM location in the eloquent cortex. This information can be useful in counseling of children and families on potential long-term neurological deficit in the setting of cerebral AVM surgery. Future study comparing long-term outcomes, both cognitive and neurological, across treatment modalities is needed.

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Disclosures

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Author Contributions

Conception and design: Ravindra, Smith. Acquisition of data: Ravindra, Eli, Lanpher, Klein. Analysis and interpretation of data: Ravindra, Eli, Lanpher, Klein. Drafting the article: Ravindra. Critically revising the article: Ravindra, Bollo, Griaudzde, Brockmeyer, Kestle, Couldwell, Scott, Smith. Reviewed submitted version of manuscript: Ravindra, Bollo, Eli, Griaudzde, Klein, Zhu, Brockmeyer, Kestle, Couldwell, Scott, Smith. Approved the final version of the manuscript on behalf of all authors: Ravindra. Statistical analysis: Ravindra, Zhu. Administrative/technical/material support: Lanpher.

Supplemental Information

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