Acute surgical management of children with ruptured brain arteriovenous malformation

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OBJECTIVE Rupture of brain arteriovenous malformation (AVM) is the main etiology of intracerebral hemorrhage (ICH) in children. Ensuing intracranial hypertension is among the modifiable prognosis factors and sometimes requires emergency hemorrhage evacuation (HE). The authors aimed to analyze variables associated with HE in children with ruptured AVM.

METHODS This study was a single-center retrospective analysis of children treated for ruptured AVM. The authors evaluated the occurrence of HE, its association with other acute surgical procedures (e.g., nidal excision, decompressive hemicraniectomy), and clinical outcome. Variables associated with each intervention were analyzed using univariable and multivariable models. Clinical outcome was assessed at 18 months using the ordinal King's Outcome Scale for Childhood Head Injury.

RESULTS A total of 104 patients were treated for 112 episodes of ruptured AVM between 2002 and 2018. In the 51 children (45.5% of cases) who underwent HE, 37 procedures were performed early (i.e., within 24 hours after initial cerebral imaging) and 14 late. Determinants of HE were a lower initial Glasgow Coma Scale score (adjusted odds ratio [aOR] 0.83, 95% CI 0.71–0.97 per point increase); higher ICH/brain volume ratio (aOR 18.6, 95% CI 13–26.5 per percent increase); superficial AVM location; and the presence of a brain herniation (aOR 3.7, 95% CI 1.3–10.4). Concurrent nidal surgery was acutely performed in 69% of Spetzler-Martin grade I–II ruptured AVMs and in 25% of Spetzler-Martin grade III lesions. Factors associated with nidal surgery were superficial AVMs, late HE, and absent alteration of consciousness at presentation. Only 8 cases required additional surgery due to intracranial hypertension. At 18 months, overall mortality was less than 4%, 58% of patients had a favorable outcome regardless of surgical intervention, and 87% were functioning independently.

CONCLUSIONS HE is a lifesaving procedure performed in approximately half of the children who suffer AVM rupture. The good overall outcome justifies intensive initial management.

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KEYWORDS arteriovenous malformation; intracerebral hemorrhage; hematoma evacuation; nidal surgery; vascular disorders

PEDIATRIC brain arteriovenous malformations (AVMs) are the main cause for intracerebral hemorrhage (ICH) in children.^{1,2} Ruptured AVM-related ICH may cause cerebral damage and result in increased intracranial pressure (ICP), leading to brain herniation, sei-

zures, neurological deficits, altered consciousness, and eventually death if not properly treated at the acute phase.

Along with critical care management, emergency surgery plays a preeminent role in reducing ICP, secondary brain injuries due to mass effect of ICH, and early re-

ABBREVIATIONS aOR = adjusted odds ratio; AVM = arteriovenous malformation; DC = decompressive hemicraniectomy; EVD = external ventricular drain; GCS = Glasgow Coma Scale; HE = hemorrhage evacuation; ICH = intracerebral hemorrhage; ICP = intracranial pressure; IVH = intraventricular hemorrhage; KOSCHI = King's Outcome Scale for Childhood Head Injury; TBV = total brain volume.

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bleeding. Acute surgical interventions include external ventricular drain (EVD) insertion, hemorrhage evacuation (HE), targeted AVM nidus excision, and, rarely, decompressive hemicraniectomy (DC).^{3–5} Yet, given the scarcity of the literature and the rarity of the condition, there are no surgical guidelines—hence, acute surgical decisions are mainly extrapolated from traumatic brain injury cases, from adults, and from local institutional experience.

In a large pediatric cohort of ruptured AVMs, we aimed to retrospectively analyze the determinants of surgical management at the acute phase, with an emphasis on HE, and their association with clinical outcome.

Methods

Study Design, Data Extraction, and Clinical Data

The manuscript was prepared in accordance with the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement.⁶ Data were retrospectively queried from a previously described prospective registry of children (age < 18 years) presenting with ICH at the pediatric hospital (Necker-Enfants Malades, Paris, France).² Children with AVM rupture between 2002 and 2018 were included in the current study.

As is customary for observational studies with retrospective analysis of routinely acquired data, generalized patient consent was used, written informed consent was waived, and patients or the children's parents could oppose the use of their health-related data for the purpose of research. Study protocol was approved by the institutional review board. Retrospective data collection was completed by studying the patients' medical charts with the use of a local data warehouse.⁷ Clinical data retrieved included age at event; sex; initial Glasgow Coma Scale (GCS) score; presence of seizures; and clinical signs of elevated ICP; as well as neurological deficits (namely motor deficits, visual field deficits, aphasia, and cerebellar syndrome). Altered mental status was considered for initial GCS score < 9. Clinical outcome, assessed 18 months after ICH, was defined as favorable if the King's Outcome Scale for Childhood Head Injury (KOSCHI) score was $\geq 5A$, as unfavorable outcome with mild disabilities for scores 4A and 4B, and as unfavorable with severe disabilities or vegetative state for KOSCHI scores 3 and 2.⁸

Imaging Analysis

The following variables were retrieved from acute imaging sequences: ICH location, ICH volume (in ml) and its ratio to the measured total brain volume (TBV; in ml) (ICH/TBV), and hydrocephalus (according to ventricular dilatation of the temporal horns with a diameter > 5 mm).⁹ The extent of the intraventricular hemorrhage (IVH) was semiquantitatively assessed using the modified Graeb score (ranging from 0 to 32), which is based on the distribution and amount of IVH and associated ventricular dilatation.¹⁰ Three types of brain herniation were distinguished: herniation of the falx, herniation of the temporal lobes, and herniation of the cerebellar tonsils. Angioarchitectural AVM characteristics included Spetzler-Martin grade, eloquent location, associated aneurysms, any deep venous drainage, and deep AVM localization. In order to

strengthen the outcome of statistical analysis given the size of our cohort, AVMs were grouped according to the classification described by Spetzler and Ponce into class A (Spetzler-Martin grade I and II), class B (Spetzler-Martin grade III), and class C (Spetzler-Martin grade IV and V).¹¹ Hemorrhages reaching the cortex or located immediately subcortically were defined as superficial.

Treatment Modalities

Because our hospital is the on-call pediatric neurosurgical department for Île-de-France region (population 12 million), children can be referred from other hospitals in case of ICH or directly from home in case of altered consciousness and/or signs of intracranial hypertension. At admission, the clinical-radiological presentation guided the choice of the initial treatment modality: a low GCS score imposed limited imaging, initial resuscitation, and discussion of emergency surgery. In the other patients, the cases were discussed in an interdisciplinary fashion, and in AVMs in which an intranidal or a flow aneurysm could be identified as the likely source of bleeding, endovascular treatment was aimed primarily at obliterating the suspect vascular segment. Conversely, if no source of bleeding was seen on baseline imaging, diagnostic catheter angiography was performed within the first week after onset.

HE Procedure

Acute surgeries were performed by more than 10 different surgeons over the study period, including junior consultants with limited experience in neurovascular surgery, highlighting the rarity of pediatric ICH. HE was performed using the following general rules. After a large craniotomy was performed under neuronavigation, the cortex was incised where the hematoma was the most superficial, while avoiding eloquent areas. Gentle aspiration of the hematoma along with irrigation with saline aimed for significant brain decompression, but not for complete clot removal so as to avoid AVM rebleeding. Simultaneous targeted resection of the AVM was performed if angioarchitectural characteristics and the surgeons' experience allowed it. If needed, an EVD was placed before or during HE. DC was performed during the initial surgery of HE in cases of uncontrollable parenchymal swelling and during a second surgical session in cases of refractory intracranial hypertension despite optimal medical treatment according to recommended guidelines.¹² HEs performed within 24 hours after initial imaging were termed "early," and those performed after 24 hours were called "late."

Statistical Analysis

Statistical analyses were performed using JMP Pro version 14.0 (SAS Institute Inc., 1989–2019). Continuous variables were expressed as the mean $\pm$ standard deviation. Categorical variables were expressed as absolute number and percentages. We tested the association of clinical and radiological variables with the occurrence of HE, using univariable tests as appropriate according to the variable's nature. Variables associated with HE that had a $p < 0.1$ were entered in a multivariable nominal logistic regression model to test their independent association with

TABLE 1. Clinical and radiological characteristics of 112 cases of ruptured brain AVMs and comparison between the group with HE and the one without

Variable	Total, n = 112	HE, n = 51	No HE, n = 61	p Value
Patient characteristics				
Age at event, yrs	9.6 ± 3.9	9.6 ± 5.8	9.7 ± 4.7	0.86
Male sex	60 (54%)	25 (49%)	35 (57%)	0.78
GCS score	11.4 ± 3.9	9.9 ± 5.6	12.7 ± 4.4	<0.001
GCS score <9	34 (30%)	23 (45%)	11 (18%)	0.002
Neurological deficit	64 (57%)	35 (69%)	29 (48%)	0.04
Seizures	83 (74%)	45 (88%)	38 (62%)	0.001
Intracranial hypertension symptoms	93 (83%)	46 (90%)	47 (77%)	0.08
Baseline imaging data				
ICH/TBV	2.2% ± 1.9%	3.2% ± 2.0%	1.4% ± 1.8%	<0.001
ICH vol, ml	21.8 ± 15.9	30.7 ± 19.3	14.4 ± 17.6	<0.001
Left-sided ICH	55 (49%)	27 (53%)	28 (46%)	0.13
Supratentorial ICH	94 (84%)	45 (88%)	49 (80%)	0.30
Modified Graeb score	9.8 ± 11.4	8.6 ± 14.4	10.8 ± 16.3	0.34
Hydrocephalus	68 (61%)	31 (61%)	37 (61%)	>0.99
Presence of brain herniation	49 (44%)	36 (71%)	13 (21%)	<0.001
Falcine herniation	28 (25%)	19 (37%)	9 (15%)	<0.001
Temporal herniation	15 (13%)	12 (24%)	3 (5%)	
Tonsillar herniation	6 (5%)	5 (10%)	1 (2%)	
No herniation	63 (56%)	15 (29%)	48 (79%)	
EVD placement	61 (54%)	26 (51%)	35 (57%)	0.57
AVM characteristics				
Superficial location	52 (46%)	32 (63%)	20 (33%)	0.002
Any deep AVM drainage	53 (47%)	18 (35%)	35 (57%)	0.02
Secondary bleeding	8 (7%)	3 (6%)	5 (8%)	0.73
Presence of AVM-associated aneurysm	36 (32%)	15 (29%)	21 (34%)	0.55
AVM eloquent cortex	60 (54%)	27 (53%)	33 (54%)	>0.99
Spetzler-Ponce class				
A	72 (64%)	37 (73%)	35 (57%)	0.02
B	26 (23%)	12 (24%)	14 (23%)	
C	14 (13%)	2 (4%)	12 (20%)	

Values are expressed as absolute number (percentage) or mean ± SD, as appropriate. Boldface type indicates statistical significance.

HE. Nonsignificant variables were then backward eliminated. Independently of univariable test results, and based on previous knowledge, it was prespecified that the multivariable model for HE would be adjusted for age, baseline ICH volume, and initial GCS score. We proceeded with the same methodology in order to test associations of clinical-radiological variables and HE with the clinical outcome. The association between HE and other surgical procedures was explored using descriptive statistics. Probability curves were calculated for HE according to initial GCS score and ICH volume as well as ICH/TBV. Two-sided p values < 0.05 were considered statistically significant.

Results

Study Sample and Baseline Characteristics

A total of 123 children with ICH due to AVM were

treated at our institution between 2002 and 2018. Among them, 6 were excluded due to missing clinical data and 13 with isolated IVH. Recurrent bleeding was observed in 8 of 104 children (7.7%); thus, a total of 112 hemorrhagic events were analyzed. Clinical and radiological findings of the entire study group, including a comparison between the group with and the group without HE, are summarized in Table 1. In brief, 60 of 112 cases (53.6%) were in males and 52 (46.4%) were in females. The mean age at rupture was 9.6 ± 3.9 years. The mean initial GCS score at admission was 11.4 ± 3.9, intracranial hypertension symptoms were reported in 93 of 112 cases (83%), a neurological deficit was noted in 64 (57.1%), and seizures in 83 cases (74.1%). Most of the ICHs (84%) were supratentorial, with a mean ICH volume of 21.8 ± 15.9 ml. Hydrocephalus and brain herniation occurred in 60.7% and 43.7% of the cases, respectively. There were 72 Spetzler-Ponce class A (64%),

FIG. 1. Chronological flow diagram of primary early HE (within 24 hours after initial cerebral imaging) and late HE (more than 24 hours after initial cerebral imaging), as well as secondary surgical interventions performed in the described cohort.

26 class B (23%), and 14 class C (13%) AVMs. The flow diagram in Fig. 1 illustrates the timeline of acute surgical interventions, which will be separately discussed below.

Hemorrhage Evacuation

In 61 cases (54.5%), children did not undergo acute surgical management of the ICH. HE was performed in 51 cases (45.5%), of which 37 cases (73%) were performed early. Due to secondary neurological deterioration, late HE was necessary in 14 cases (mean time interval to initial cerebral imaging 3.3 days, range 1–12 days). In 7 cases, catheter angiography was performed before HE, including 2 cases with presurgical embolization. In 2 cases, concur-

rent DC was required because of uncontrolled brain swelling. There was a significant AVM rupture during HE in 1 case that could eventually be controlled after extensive AVM coagulation.

In univariate analysis, patients with HE had a lower initial GCS score than patients without HE (9.9 vs 12.7, $p < 0.001$), more often had neurological deficits (69% vs 48%, $p = 0.04$), more commonly had seizures (88% vs 62%, $p = 0.001$), had more frequent brain herniation on initial cerebral imaging (73% vs 27%, $p < 0.001$), had larger ICH volumes (30.7 ± 19.3 ml vs 14.4 ± 17.6 ml, $p < 0.001$), and had higher ICH/TBV ratios ($3.2\% \pm 2.0\%$ vs $1.4\% \pm 1.8\%$, $p < 0.001$). The underlying AVM was more frequently superficial (63% vs 33%), with lower Spetzler-Ponce class

TABLE 2. Multivariable model analysis of predictive factors for HE after ruptured brain AVM

Model 1			Model 2		
Variable	aOR (95% CI)	p Value	Variable	aOR (95% CI)	p Value
GCS score, per point increase	0.83 (0.71–0.97)	<0.001	GCS score, per point increase	0.84 (0.71–0.99)	0.04
Age at event, per yr increase	0.97 (0.85–1.11)	0.67	Age at event, per yr increase	0.99 (0.86–1.13)	0.86
ICH/TBV, per % increase	18.6 (13–26.5)	0.01	ICH/TBV, per % increase	21.2 (14.2–31.8)	<0.001
Superficial AVM	3.75 (1.25–11.31)	0.02	Superficial AVM	3.57 (1.14–11.22)	0.03
Presence of brain herniation	3.73 (1.34–10.39)	<0.001	Falcine herniation*	2.04 (0.61–6.79)	0.24
			Temporal herniation*	7.71 (1.34–44.32)	0.02
			Tonsillar herniation*	14.3 (1.28–159.98)	0.001

Boldface type indicates statistical significance.

* "No herniation" used as reference.

(97% class A vs 70% class B) (Table 1). In the fully adjusted multivariable model, factors independently associated with HE included high ICH/TBV (adjusted odds ratio [aOR] 18.6, 95% CI 13.0–26.5 for each percent increase; $p = 0.01$), low initial GCS score, presence of brain herniation, and superficial AVM location (aOR 3.75, 95% CI 1.25–11.3; $p = 0.02$) (Table 2). When considering the subtype of brain herniation, tonsillar herniation was most strongly associated with HE (aOR 14.3, 95% CI 1.28–159.98; $p = 0.001$). Figure 2 is a graphical representation of the adjusted multivariable analysis (model 1 in Table 2) probability of HE plotted against initial GCS score and ICH volume.

Compared with the group undergoing late HE ($n = 14$ cases), the group with early HE ($n = 37$ cases) had lower initial GCS scores (8.6 ± 0.6 vs 13.2 ± 0.9 , $p < 0.001$), more frequent altered consciousness (59% vs 7%, $p < 0.001$), a more infratentorial location (16.2% vs 0%, $p = 0.002$), and a more frequent presence of brain herniation on initial

cerebral imaging (81.1% vs 42.9%, $p = 0.01$). Conversely, no significant difference of ICH/TBV was found ($3.2\% \pm 1.4\%$ vs $2.9\% \pm 1.4\%$, $p = 0.79$). Predictive factors for the early HE timing included low initial GCS score, infratentorial AVM, and the presence of brain herniation on initial cerebral imaging.

AVM Nidal Resection at the Acute Phase

Twenty-one of 51 patients (41%) had concurrent AVM nidus resection and HE (12 during early HE and 9 during late HE) (Fig. 3). Resected AVMs were either Spetzler-Ponce class A (7 Spetzler-Martin grade I and 11 Spetzler-Martin grade II) or B (3 Spetzler-Martin grade III). Hence, nidus surgery was performed in 69% of Spetzler-Ponce class A AVMs and 25% of Spetzler-Ponce class B AVMs undergoing HE. Factors associated with concurrent nidus resection, compared with exclusive HE, were higher initial GCS scores (mean 12.95 ± 2.5 vs 8.6 ± 3.7 , $p < 0.001$) and

FIG. 2. Probability of HE in the fully adjusted model, plotted according to the initial GCS score (A) and ICH volume (B). Figure is available in color online only.

FIG. 3. Sankey flow diagram of surgical interventions. Illustration of the chronology of surgical interventions according to the Spetzler-Ponce grade of the ruptured AVMs: Spetzler-Ponce class A (left, Spetzler-Martin grade I–II), class B (center, Spetzler-Martin grade III), and class C (right, Spetzler-Martin grade IV–V). The widths of the bands represent the proportion of cases per group undergoing early or late surgery as well as targeted nidus resection. S M Grade = Spetzler-Martin grade. Figure is available in color online only.

accordingly less frequent altered consciousness (5% vs 63.6%, $p < 0.001$), more frequent superficial AVMs (92.3% vs 52.6%, $p = 0.02$), and a late HE (47.6% vs 8%, $p = 0.03$). There was a tendency toward lower Spetzler-Ponce classes in the AVM resection group (85.7% Spetzler-Ponce class A vs 65.3%, $p = 0.07$). Complete resection was attempted in 9 of 21 cases (43%). Five underwent diagnostic catheter angiography before surgery, and angioarchitectural characteristics were evaluated with injected CT scans in the remainder.

Surgery for Secondary Intracranial Hypertension

Five patients underwent secondary decompressive craniectomy because of refractory intracranial hypertension despite optimal medical treatment, with a mean delay of 3.5 ± 4.4 days after admission. These patients had lower initial GCS scores (7.1 ± 3.7 vs 11.8 ± 3.7 , $p = 0.02$), larger ICH volumes (39.0 ± 16.7 ml vs 20.5 ± 15.4 ml, $p = 0.02$), a larger ICH/TBV ($4.1\% \pm 2.0\%$ vs $2.1\% \pm 1.6\%$, $p = 0.02$), and more frequent brain herniation on initial cerebral imaging than patients who did not require DC.

Outcome

During the acute phase, rebleeding occurred in 2 cases, after a delay of 2 and 7 days since admission for the first bleeding episode. One of the patients, with a Spetzler-Martin grade II AVM, had undergone early HE with concurrent partial AVM resection and underwent successful reoperation at day 7; the other, with a Spetzler-Martin grade IV AVM, died after 48 hours.

Postoperative complications requiring additional surgery were encountered in 2 cases. One patient underwent reoperation because of wound healing difficulties, and the other patient required enlargement of the craniectomy due to recurrent intracranial hypertension.

At 18 months, the clinical outcome was available for 101 of 104 patients, and it was pending for the 3 remaining children. Distribution according to the KOSCHI scale was as follows for these 101 patients: KOSCHI 5B in 21 cases (20.8%); KOSCHI 5A in 38 (37.6%); KOSCHI 4B in 23 (22.8%); KOSCHI 4A in 6 (5.9%); KOSCHI 3 in 8

(7.9%); KOSCHI 2 in 1 (1%); and 4 patients (4%) died. In summary, 58.4% of the patients had a favorable outcome (KOSCHI 5A and 5B); 28.7% had an unfavorable outcome with mild disabilities yet functioning independently (KOSCHI 4A and 4B); 8.9% had severe disabilities or were in a vegetative state (KOSCHI 3 and 2); and 4% died. Hence, 87% were functioning independently 18 months after the bleeding event.

There was no significant difference between patients with and those without HE concerning favorable outcome (KOSCHI $< 5A$, 52% vs 33%; $p = 0.05$) and severe disability (KOSCHI $< 4A$, 18% vs 8%; $p = 0.16$). There was no significant difference either between patients undergoing early HE compared to those with late HE (KOSCHI $< 5A$, 60% vs 33%, respectively; $p = 0.07$) or between patients with late HE compared to those with no HE required (KOSCHI $< 5A$, 33% vs 31%, respectively; $p = 0.13$).

Discussion

Compiling more than 110 pediatric ruptured brain AVM cases, from the hyperacute phase to 18 months after stroke onset, we showed that a surgical strategy comprising HE in nearly half of children is associated with functional independence in approximately 87% and a mortality rate as low as 3.8%. Interestingly, outcome was comparable between surgically and nonsurgically treated patients despite significantly different initial clinical presentation. In our opinion, these results reinforce an interventionist surgical strategy, based on the severity of initial clinical status as well as baseline imaging, including brain herniation and large ICH volumes.

Previous reports on emergency surgery in children with ruptured AVMs are scarce. In Nerva et al.,¹³ between 2005 and 2012, 16 of 27 patients (59%) underwent nonelective—thus emergency or urgent—surgery after a mean delay of 4 days between admission and surgery. This included 3 cases of primary DC and 2 cases of secondary DC due to postoperative cerebral edema.¹³ Shtaya et al. reported, over a period of more than 20 years (1994–2016), HE in 5 of 37 children (14%) during the initial hospitalization for ruptured brain AVM.¹⁴ Singhal et al. described HE in 12

of 15 children (80%) presenting with a ruptured AVM and an initial GCS score of < 9 , including nidus resection in 9 cases, with a median 4-hour interval after admission and a mortality rate of 20%.¹⁵

When comparing our results to adult cohorts of patients with ruptured AVM, the most striking difference is the higher frequency of HE, as well as the shorter time interval between initial cerebral imaging and surgery.^{16,17} Beecher et al. described emergency surgery in only 6 of 163 cases with ruptured AVM.¹⁸ In another adult cohort of 59 patients from Finland, the median delay for surgery was 2 days after onset of symptoms.¹⁹ Both differences are striking because the mean ICH volume was not higher in the present study than in adults when taking into account whole-brain volume (mean ICH/TBV 2.2%). Indeed, 60 ml is frequently used as a cutoff value for surgery in adults and is comparable to a mean ICH/TBV of 4% in a child. Rapid decompensation of intracranial hypertension in children may be explained by differences in brain parenchyma compliance,²⁰ fewer CSF-related compensatory mechanisms, and a higher rate of seizures. Altogether, despite the fact that the present study may reflect a single institution's habits, the large proportion of early HE in this large sample strongly supports the immediate transfer of children after ICH diagnosis to a tertiary care center with a pediatric neurointensive care unit and neurosurgical facilities.

Identifying determinants of surgical HE is important, because these emergency procedures are often performed in the off-hours by surgeons with various levels of neurovascular experience.¹⁹ Yet, few data exist on these specific procedures, and there are no evidence-based recommendations, because both children and underlying vascular lesions were excluded from randomized controlled trials investigating the benefits of HE (such as Surgical Trial in Intracerebral Hemorrhage [STICH] I or II).^{21,22} Experts advocate ICH evacuation in cases of "large, life-threatening hematoma"²³ or in cases of "decreased level of consciousness due to an ICH" and for "hematomas larger than 30 ml in the temporal lobe or cerebellum or 60 ml in other parts of the hemispheres."²⁴ In the present study, HE was more frequently performed in children with altered consciousness, larger hematoma, or radiological signs of herniation, which emphasizes the need for a global clinical and imaging evaluation for the surgical indication. Although the sample size of our cohort did not allow us to derive and validate a clinical-radiological score to guide surgical indications, we supply probability curves, which may help in decision-making, notably for children with initial GCS scores > 10 and intermediate ICH volumes. As a representative example, in a 10-year-old child with an initial GCS score of 11 and a superficial ruptured AVM, the probabilities of evacuating the ICH with ICH/TBV rates of 0.5%, 2%, and 5% would be 25.9%, 46%, and 84%, respectively.

Whereas there is no ambiguity regarding the indication for emergency surgery for a child who presents in extremis, the timing of nidus surgical treatment for ruptured AVMs has been a controversial topic. In the absence of a recommended timeline, timing is a subject of significant variation in clinical practice, between AVM resection concurrent with lifesaving emergency craniotomy and clot

removal within a few hours after onset versus delayed for weeks.¹⁸ Interestingly, in the present study, 21 of 51 patients (41%) had concurrent HE and AVM nidus resection, in line with previous series reporting from 20%¹⁴ to 84%²² early AVM removal. The attitude in favor of delayed nidus resection originated from concerns about compression of the nidus by the hematoma, hindering the visualization of the AVM architecture and potentially damaging the brain tissue. Therefore, early AVM surgery can increase both the risk of an incomplete removal²⁴ and the risk of neurological deficit.^{23,25} The fact that AVMs are unlikely to rebleed very early has also been used to support this opinion. However, from another neurosurgical perspective, fresh AVM-related hematoma may facilitate resection, creating a cavity and offering a good cleavage plane. This could explain why in the present series AVM resection was more frequently performed after 24 hours in patients with a higher GCS score and more superficial AVM location.

In addition, immediate resection prevents a new hemorrhage and facilitates the rehabilitation of the patient. Hence, some authors advocate concurrent HE and microsurgical resection in a single procedure at the acute phase, notably for low Spetzler-Martin grade AVM, with comparable outcomes to delayed surgery and a reduction of the rebleeding risk.^{16,19,26} However, in the present study we did not observe an increased risk of rebleeding in children with delayed AVM excision and did not find such reports in the pediatric literature. We encountered a rebleeding rate of 7.7%, with only 2 cases of early rebleeding during initial hospitalization. This rate is comparable to the rebleeding rate of 6%–15.6% within the first year after AVM rupture reported in adult series.²⁷ Combined endovascular and microsurgical procedures with preoperative embolization is proposed even at the acute stage because it increases the obliteration rate of AVM even for grade III lesions and decreases intraoperative risk.^{28,29} Yet, this solution may not appear ideal for children with ICH and severe clinical presentation (e.g., coma, third nerve palsy) who require emergency surgery without the time for extensive imaging³⁰ or neurointerventional treatment.

It is important to note that, despite the high rate of emergency neurosurgical procedures in our sample, the overall mortality rate (3.8%) was low, and lower than in previous reports in this age group after AVM rupture (ranging between 5% and 20%).^{13,31–33} Functional independence was observed in almost 9 in 10 children at 18 months. Previous studies reported similar independent functioning in 83%–92% and favorable outcome in 61%–84% of surgically treated ruptured AVMs in children, but with shorter-term follow-up.^{13,15,34} Long-term outcomes for children with early HE were comparable to those with delayed HE, and similar between children with delayed HE and those without HE. This is despite the fact that the patients with life-threatening hematomas, and therefore disproportionately more severe initial presentations, were naturally treated with minimal delays. These results reinforce the role of acute neurosurgical decompressive approaches in determining clinical outcome.

Study Strengths and Limitations

Our study has several strengths, including a large cohort

of consecutively treated children, long-term follow-up, and use of a multivariate model aiming to enable the analysis of the time interval between ICH onset and surgery as a potential predicting factor with regard to outcome. Limitations included the retrospective design and the inclusion of patients with infratentorial AVMs, because infratentorial ICH often necessitates immediate surgery, and timing of treatment is less controversial. Also, one may argue that too many surgeons with various experience in pediatric neurovascular surgery were involved, which may have maximized surgeon experience as a confounding factor. However, in our opinion, even though AVM neurosurgical management is a difficult task, our study offers real-world care conditions with generalizable results. Prospective investigations involving a larger number of centers are required to validate the findings of this study.

Conclusions

The HE rate in children with ruptured brain AVM is high and is associated with clinical and imaging baseline severity. With almost half of children requiring HE in this study, the mortality rate was limited to less than 4%, supporting an intensive approach for children with severe presentations. In addition, neither the time interval between AVM bleeding and HE nor the HE itself negatively influenced clinical outcome, with functional independence observed in 9 of 10 children at 18 months. The advantage of concurrent acute nidal resection is balanced between the decreased risk of early rebleeding and the elevated risk of increased cerebral damage.

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Disclosures

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

Author Contributions

Conception and design: Blauwblomme, Stricker, Boulouis, Benichi, Naggara. Acquisition of data: Blauwblomme, Stricker, Boulouis, Benichi, Gariel, Garzelli, Hak, Alias, Kerleroux, Beccaria, Chivet, de Saint Denis, James, Paternoster, Zerah, Kossorotoff, Boddaert, Brunelle, Meyer, Puget, Naggara. Analysis and interpretation of data: Blauwblomme, Stricker, Boulouis, Benichi, Bourgeois, Naggara. Drafting the article: Blauwblomme, Stricker, Boulouis, Naggara. Critically revising the article: Blauwblomme, Stricker, Boulouis, Benichi, Naggara. Reviewed submitted version of manuscript: Blauwblomme, Stricker, Boulouis, Benichi, Naggara. Approved the final version of the manuscript on behalf of all authors: Blauwblomme. Statistical analysis: Stricker, Boulouis. Administrative/technical/material support: Stricker, Boulouis. Study supervision: Blauwblomme, Benichi, Naggara.

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