Emergency management of deeply comatose children with acute rupture of cerebral arteriovenous malformations

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Purpose: To assess the impact of emergency management on mortality and morbidity of acute rupture of cerebral arteriovenous malformations resulting in deep coma in children, and the factors predicting outcome.

Methods: Retrospective chart review of 20 children with a Glasgow Coma Scale ≤ 8 with acute hemorrhagic stroke from a cerebral arteriovenous malformation rupture was conducted. Protocol included: early resuscitation with tracheal intubation and ventilation after induction of anesthesia with sufentanil, and benzodiazepine, and mannitol 20% or hypertonic saline 7.5% infusion for life-threatening brain herniation. Radiological exploration was limited to contrast-enhanced CT scan preceding immediate surgical decompression. Postoperatively, children were deeply sedated and intracranial pressure monitoring allowed titration with osmotherapy, vasopressors, hyperventilation or barbiturate coma to control cerebral perfusion pressure. Analysis used stratification of the type of hemorrhage (supra or infra tentorial), location (intraparenchymal and subarachnoid, intraparenchymal and intraventricular or intraventricular alone) and relationship between presentation, evolution with resuscitation, type of cerebral lesion, and outcome.

Results: Patients had a severe initial presentation (median Glasgow Coma Scale five), eight had unilateral and eight bilateral third nerve palsy. Compressive hematoma in supratentorial localisation represented 75% of the cases. Global mortality was 40%. Persistence of mydriasis after resuscitation increased mortality to 75%. Massive intraventricular flooding was associated with increased mortality. Good functional outcome was achieved in survivors.

Conclusion: Acute rupture of an AVM can result in rapidly progressing coma. Emergency management with early resuscitation, minimal radiological exploration before rapid surgical decompression results in a mortality rate of 40%, but a good functional outcome can be expected in the survivors.

Objectif : Évaluer l'effet et le pronostic d'une intervention d'urgence sur la mortalité et la morbidité d'une rupture imprévisible de malformations artérioveineuses cérébrales qui ont conduit à un coma profond chez des enfants.

Méthode : Un examen rétrospectif des dossiers de 20 enfants, cotés ≤ 8 à l'échelle de Glasgow, qui ont subi un accident hémorragique aigu causé par la rupture de malformations artérioveineuses cérébrales, a été réalisé. Le protocole comportait : une réanimation précoce, une intubation endotrachéale et une ventilation après l'induction de l'anesthésie avec du sufentanil, du mannitol à 20 % ou une perfusion salée hypertonique à 7,5 % pour traiter l'hernie cérébrale grave. L'exploration radiologique s'est limitée à la scanographie de prise de contraste juste avant la décompression chirurgicale. Après l'opération, les enfants ont reçu une forte sédation. Le monitoring de la pression intracrânienne a permis le titrage de l'osmothérapie, des vasopresseurs, de l'hyperventilation ou du coma barbiturique pour contrôler la pression de perfusion. L'analyse a porté sur la stratification du type d'hémorragie (sus-tentorielle ou sous-tentorielle), la localisation (intraparenchymateuse et sous-arachnoïdienne, intraparenchymateuse et intraventriculaire ou intraventriculaire seulement) et la relation entre la condition initiale, l'évolution de la réanimation, le type de lésion cérébrale et les résultats.

Résultats: Les patients présentaient une condition initiale sévère (cinq à l'échelle de Glasgow), huit avaient une paralysie unilatérale du troisième nerf et huit, bilatérale. L'hématome compressif sus-tentorial représentait 75 % des cas. La mortalité globale a été de 40 %. La persistance de mydriase après la réanimation a fait augmenter la mortalité à 75 %. L'épanchement intraventriculaire massif a été associé à un accroissement de la mortalité. Une bonne évolution fonctionnelle a été possible chez les survivants.

Conclusion : La rupture aiguë de MAV peut rapidement évoluer vers le coma. Une intervention d'urgence par une réanimation précoce et une exploration radiologique minimale avant la décompression chirurgicale rapide ont

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Accepted for publication April 30, 2000.

CEREBRAL arteriovenous malformations are diagnosed during childhood in less than 20% of the cases reported in the literature.¹ The first manifestation, in 60 to 80% of these pediatric cases, is an abrupt rupture resulting in intracranial hemorrhage.²⁻⁴ Although most series reporting AVMs in children include some cases of acute rupture resulting in compressive hematoma and massive intraventricular flooding with deep coma,⁵ this particular entity and its specific emergency management have not yet been individualised. The aim of this study is to report our experience of acute management of AVM rupture resulting in deep coma. The results on mortality and morbidity of a therapeutic protocol including emergent aggressive critical care, minimal radiological exploration and early surgical decompression were analysed.

Patients and methods

Patients inclusion

This study was approved by the review board of our department who stated that informed consent for this chart review study was not required. Over 18 months, the records of 40 children admitted to our institution with the diagnosis of acute rupture of a cerebral AVM were analysed. Only those who had a Glasgow coma score (GCS) of eight or less at the time of the actual bleeding were included in this study. The review consisted in an analysis of the recent medical history including headaches, seizures, nausea and vomiting, or the abrupt occurrence of a focal neurological deficit. The clinical condition at the moment of the initial medical evaluation and the progression of the neurological status were noted.

Initial management

A specific management protocol has been designed for these particular neuroemergencies. It was initiated either outside the hospital by physicians of mobile intensive care units (SAMU) or by our team in children who deteriorated during their hospital course. It included emergency tracheal intubation (ETI) and moderate hyperventilation after intravenous induction of general anesthesia in every comatose child. Immediately before emergency CT scan exploration, catheters for continuous arterial and central venous pressure monitoring were rapidly inserted. Osmotherapy with mannitol 20% in 0.5 g·kg⁻¹ bolus or rapid infusion of 3 ml·kg⁻¹ hypertonic saline 7.5% solution were used when evidence of brain herniation was present. The target range for plasma osmolality was 300-310 mosmol·kg.

Radiological evaluation

Emergency exploration was limited to a contrast-enhanced spiral CT-scan examination within the first two hours following the episode of acute deterioration. Cerebral angiography was not undertaken before emergency surgical decompression in these very severely ill children. On this basis, suspected ruptured AVMs were classified into three groups with reference to the extension of the hemorrhage. Group A included children with parenchymal hematoma and rupture into the subarachnoid space, group B children with a hematoma ruptured into the ventricle, and group C children presenting with isolated intraventricular hemorrhage. Diagnosis of the anatomic type of AVM was made either upon operative findings or upon the results of delayed cerebral angiography on those children who survived.

Surgical management

Emergency surgical operations were performed whatever the clinical status after CT scan. Simple external ventricular drainage was performed when ventricular hemorrhage was isolated. It was associated with a surgical resection of the intraparenchymal hematoma and cautious micro-coagulation when a compressive hematoma resulting in acute midline shift was present. Only those AVMs that were superficial enough to be completely exposed after brain decompression were resected microsurgically at the first operation. In others, no attempt was made at this stage for complete resection of the AVM. At the end of the operation, decompressive craniotomy and duralplasty with a patch were performed when direct dural closure was not possible.

Postoperative management

All patients were maintained under general anesthesia with a continuous infusion of sufentanil (0.2-0.5 g·kg⁻¹·hr⁻¹) and flunitrazepam (0.01-0.03 mg·kg⁻¹·hr⁻¹). Normothermia was maintained and paracetamol (150 mg·kg⁻¹·day⁻¹ *iv*) was used systematically in order to avoid hyperthermia. Controlled ventilation was adjusted to maintain normoxia and normocarbida. Intracranial pressure (ICP) was monitored by means of an intraparenchymal electronic transducer (Codman© Neurotrend ICP monitor). When needed, a continuous infusion of dopamine was used to improve mean arterial blood pressure and to preserve cerebral perfusion pressure (CPP) > 60 mmHg. In the case of a persistent raised ICP, a retrograde jugular bulb fibreoptic catheter was used to monitor jugular bulb oxygen saturation (SvJO₂) continuously before therapeutic hyperventilation could be instituted. This

kind of monitoring allowed titration of hyperventilation according to cerebral hemodynamic conditions. As a last resort, pentobarbital was used in $5 \text{ mg} \cdot \text{kg}^{-1}$ bolus followed by a continuous infusion of 3 to $5 \text{ mg} \cdot \text{kg}^{-1} \cdot \text{hr}^{-1}$ when all other therapeutic maneuvers had failed to control ICP. Pentobarbital was then titrated to obtain burst suppression on EEG. All survivors had a cerebral angiography when their neurological status had improved. When an AVM was still present at this time, further management consisted of endovascular treatment with embolization whenever possible. In children with residual AVM, a delayed surgical resection was scheduled when the residual AVM was accessible to surgery. In other cases, a radiosurgical treatment was proposed.

Final outcome was assessed at the end of the acute phase and at least six months after the initiation of neurological rehabilitation in a specialised centre. Analysis of focal neurological deficit, residual epilepsy,

and Glasgow Outcome Score (GOS) to assess global functional outcome were used.⁶ This score includes five categories: a good functional result referred to good recovery or moderate persistent disability and poor results to permanent severe disability, vegetative state, or death.

Analysis of factors possibly affecting mortality, immediate outcome, and outcome at six months used Mann Whitney U-test and χ^2 analysis as appropriate. Statistical significance was considered with $P < 0.05$.

Results

The charts of 40 children hospitalised between March 1996 and December 1997 with the diagnosis of ruptured AVM were reviewed. Among them, 20 had a $\text{GCS} \leq 8$ and were selected for study. Age at presentation was $9.7 \pm 3.3 \text{ yr}$ (median 9.5, range 2-15). There was no child under one year of age, and no Gallen vein malformations. There were eight females and 12 males. The previous recent medical history of the children is presented in Table I. Only one child had a previously diagnosed AVM treated with partial embolization and experienced a new bleeding episode one year later.

Clinical presentation at the initial medical evaluation and evolution of the symptoms is listed in Table II. At the first medical evaluation, the median GCS was nine (range 3-14), a focal neurological deficit was

TABLE I Recent medical history before acute stroke

Symptoms	N	%
Headache	13	65
Nausea and vomiting	6	30
Seizures	1	5
Abrupt neurological deficit	2	10
None	5	25

TABLE II Characteristics of the 20 children upon medical evaluation and evolution of the neurological presentation

Patient	GCS 1	Motor response 1	3rd nerve palsy	Time of onset	Prehospital management	Evolution	GCS 2	Motor response 2	3rd nerve palsy
1 Ple.	4	Deposturing	Left	Abrupt	ETI+VA+IVA	Deterioration	4	Decerebrate	Bilateral
2 Pet.	10	Right hemiparesia	None	Abrupt	none	Deterioration	4	Decerebrate	Bilateral
3 Zie.	5	Deposturing	None	3 hr	ETI+VA+IVA	Deterioration	4	Decerebrate	Bilateral
4 Mit.	12	Left hemiparesia	Left	Abrupt	none	Deterioration	7	Localize	Left
5 Neg.	12	Normal	None	Day 1	none	In hospital	8	Withdraw	Left
6 Bou.	13	Left hemiparesia	None	Day 1	none	In hospital	7	Localize	Left
7 Ped.	8	Right hemiparesia	None	Abrupt	ETI+VA+IVA	Deterioration	5	Deposturate	None
8 Rah.	12	Left hemiparesia	None	Week	none	Deterioration	4	Decerebrate	Bilateral
9 Des.	14	Normal	None	Day 2	none	In hospital	4	Decerebrate	Left
10 Khe.	3	None	None	Abrupt	ETI+VA+IVA	Improvement	6	Localize	None
11 San.	9	Localize	None	Day 1	none	In hospital	8	Localize	Right
12 Dos.	6	Deposturing	Right	Abrupt	ETI+VA+IVA	In hospital	4	Decerebrate	Bilateral
13 Bar.	5	Deposturing	Right	Day 1	ETI+VA+IVA	In hospital	4	Decerebrate	Bilateral
14 Lar.	14	Normal	None	Day 3	none	Post embolization	8	Left hemiparesia	None
15 Cal.	4	Decerebrate	Left	Day 1	ETI+VA+IVA	Deterioration	3	None	Bilateral
16 Dom.	8	Normal	None	Abrupt	ETI+VA+IVA	Deterioration	3	None	Bilateral
17 Ber.	12	Normal	None	Week	none	In hospital	7	Withdraw	Left
18 Mer.	6	Left hemiparesia	None	Day 1	ETI+VA+IVA	In hospital	5	Deposturate	None
19 Mon.	8	Left hemiparesia	None	Abrupt	ETI+VA+IVA	Deterioration	5	Deposturate	Left
20 Les.	8	Normal	None	Day 1	ETI+VA+IVA	Deterioration	5	Deposturate	Right

ETI: Emergency tracheal Intubation; AV: Assisted Ventilation; IVA: Intravenous Anesthesia; In hospital: Deterioration in the hospital

TABLE III Initial resuscitation manoeuvres in the hospital before surgical decompression

<i>Resuscitation manoeuvres</i>	<i>n (%)</i>
Medical treatment	20 (100)
- Assisted ventilation	20 (100)
- <i>iv</i> sedation	19 (95)
- Hypertonic saline infusion	7 (35)
- Mannitol 20%	13 (65)
- Dopamine infusion	12 (60)
Invasive monitoring	20 (100)
Ventricular drainage	2 (10)

TABLE IV Emergency CT-scan findings

<i>Type of hemorrhage</i> <i>n (%)</i>	<i>Type of hematoma</i> <i>n (%)</i>
Group A: 8 (40)	None: 2 (10)
Group B: 10 (50)	Temporo-parietal: 6 (30)
Group C: 2 (10)	Posterior fossa: 5 (25)
	Deep: 5 (25)
	Other: 2 (10)

* One frontal, one occipital

present in seven children, four children had ipsilateral third nerve palsy and none had bilateral fixed dilated pupils. Fifteen children experienced an acute onset outside the hospital, 11 had a GCS ≤ 8 at initial presentation, were intubated at the scene, and rapidly transported to our centre. Another subset of four patients who were not comatose at the first on-scene evaluation deteriorated acutely and became comatose during transportation. None of the 15 patients who became comatose before hospital admission died during transportation, but neurological deterioration was noted in all but one patient (median GCS: 5, range 3-6 upon admission). Five children deteriorated after they had been initially admitted to our critical care unit in less dramatic medical condition with a diagnosis of acute rupture of AVM. One of these five children did so immediately after embolization of a thalamic AVM. At the moment of the in-hospital emergent critical care management, eight children had bilateral fixed pupils and eight had ipsilateral third nerve palsy, seven patients decerebrated and five had evidence of decortication. Initial resuscitation manoeuvres are summarised in Table III. They resulted in at least transient stabilisation of the neurological status in all the children and regression of pupillary dilatation before operation in 10 out of the 16 children (62%) presenting with pupillary abnormalities.

TABLE V Angiographic explorations and timing of realisation

<i>Patient</i>	<i>Angiographic findings</i>	<i>Embolization</i>	<i>Timing of angiography</i>
2	Anterior choroidal	Yes	Day 10
3	PICA	Yes	Day 10
4	Normal	No	Day 5
6	Right internal temporal	Delayed	Day 1 & 6
7	Normal	No	Day 5
9	Right internal temporal	No	Day 6
10	Left PICA and PSCA	No	Day 1
12	Normal	No	Day 5
14	Right PICA	Yes	Day 1
15	Left MCA	Yes	Day 6
16	Right PICA and PSCA	No	Day 15
18	Left choroidal pedicles	No	Day 1
19	Left callosal marginal	No	Day 1
20	Normal	No	Day 5

PICA: Posterior and inferior cerebellar artery; MCA: Middle cerebellar artery; PSCA: Posterior and superior cerebellar artery.

TABLE VI Final outcome

	<i>GOS at discharge</i> <i>n (%)</i>	<i>GOS 6 mo</i> <i>n (%)</i>	<i>GOS 12 mo</i> <i>n (%)</i>
Bad outcome	15 (75)	13 (65)	9 (45)
- Death (5)	8 (40)	8 (40)	8 (40)
- vegetative state (4)	0	0	0
- Severe disability (3)	7 (35)	5 (25)	1 (5)
Good outcome	5 (25)	7 (35)	11 (55)
- Moderate disability (2)	5 (25)	4 (20)	6 (30)
- Good recovery (1)	0	3 (15)	5 (25)

All the children had a contrast-enhanced CT scan that demonstrated an intracranial hemorrhage. The results of the neuroradiological explorations are summarised in Table IV. In spite of contrast-enhanced examination and 3-D reconstruction, the preoperative emergency explorations did not evidence the existing AVM in any case. Immediately after CT-scan evaluation, emergency surgical intervention consisted in a simple external ventricular drainage as the first operation in 10 children, with evacuation of the hematoma as the second operation in seven, evacuation of the hematoma and concomitant partial resection of a superficial AVM during the same session in five, and simple evacuation of the hematoma during the first session in five children. Dural plasty was performed in five children and decompressive craniotomy was needed in three. Twelve children experienced objective preoperative arterial spasm and were started on continuous nimodipine infusion. Three children (15%) died during or immediately after surgery.

The survivors were maintained under general anesthesia and mechanically ventilated at least for the first two postoperative days. The mean duration of mechanical ventilation and ICU stay were 7 ± 7 days (range: 2-29) and 11 ± 6 days. The initial mean value of ICP was 15 ± 13 mmHg (range: 9-50) and the mean maximal value was 34 ± 20 mmHg (range: 9-80). Plasma osmolality was carefully maintained within the range of 300 to 310 mosmol·kg⁻¹ during the first 48 hr after surgery. An infusion of mannitol or hypertonic saline solution was required at some time in the postoperative period to control ICP in all the children. Hemodynamic support with a continuous infusion of dopamine was added in 12 children. Recombinant tissue plasminogen activator (rTpa) was injected in the ventricular drainage to maintain its patency in two patients. Transcranial Doppler examination was performed systematically in all the children within six after completion of surgery. In the twelve children which experienced objective preoperative vasospasm, it depicted slightly increased mean blood flow velocities with an increased middle cerebral to carotid artery gradient evidencing possible vasospasm. Postoperative infusion of nimodipine was used in these twelve patients. In five children with untractable raised ICP, a retrograde fiberoptic jugular vein catheter was added. It demonstrated in all of them a cerebral hyperemia commanding severe hyperventilation to control cerebral perfusion pressure, and one patient required barbiturate coma.

Cerebral angiography was performed in five conscious hospitalised children before neurological deterioration had occurred. Only one child had a partial embolization of a thalamic AVM in the same session but deteriorated and became comatose immediately after. A delayed postoperative cerebral angiography was performed in nine of the patients whose neurological status improved. The results and timing of angiographic examinations are presented in Table V. Endovascular treatment of the remaining AVM was performed in five children, two children had a single session for embolization, and three required multiple sessions. A secondary radio-surgical treatment was performed in one child with a complex posterior fossa AVM after partial surgical excision and subsequent embolization.

The overall mortality was 40% of the cases including a 15% perioperative mortality. There was no increased mortality in younger children. The incidence of death was higher in those children who presented immediately before surgical intervention with ipsilateral third nerve palsy and bilateral fixed pupils in spite of aggressive resuscitative management (33% and 75% mortality

vs 0% in children with normal pupils, $P < .04$). There was also a trend ($P = .05$) toward an increased mortality in children experiencing massive intraventricular hemorrhage (20% mortality in group A, 50% in group B and 100% in group C). Neither the GCS just before operation, its motor component, nor the precise location of the AVM correlated with mortality. Interestingly, there was no survivor in a persistent vegetative state upon discharge, but seven children had severe disability at that time. After six months of intensive neurological rehabilitation, the Glasgow Outcome Score improved slightly and 12 mo after discharge, five children had no serious disability and six had moderate disability mainly represented by an ipsilateral hemiparesia (Table VI). There was no residual epilepsy requiring long-term treatment nor persistent hydrocephalus requiring permanent CSF shunting.

Discussion

In this series, deeply comatose children represented 50% of the patients presenting with an acute rupture of a cerebral AVM. This large proportion could be explained by the fact that children with clinical evidence of acute non traumatic neurological accidents were systematically managed outside the hospital by a critical care medical team insuring their immediate survival and dispatched directly to a single regional centre. Critical care management including tracheal intubation (ETI) and initiation of controlled ventilation were performed either outside or upon arrival to the hospital. Basic critical care management gained the time sufficient for performing minimal radiological explorations before immediate surgical decompression. In patients presenting with pupillary abnormalities upon admission, this resuscitation resulted in regression of pupillary abnormalities in 62% of the cases. The immediate efficiency of critical care management upon neurological symptoms was determinant for final survival, since 75% of the children who had persistent bilateral mydriasis after resuscitation died. Immediate surgical decompression performed in patients with brain herniation, or brainstem compression resulted in a 15% perioperative mortality. In the early postoperative period, control of CPP needed intervention in most children. Brain swelling was noted in five out of 17 survivors. Deep hyperventilation was effective in reducing cerebral blood flow and in adapting it to cerebral metabolism in four of the children. Vasospasm was another concern since 12 patients experienced perioperative spasm that was still present, despite continuous nimodipine infusion, at postoperative transcranial Doppler examination. A global 40% mortality was noted. A good outcome was noted 12 mo after the initial accident in 11 of the 12 survivors.

Among these patients, six had a persistent moderate disability and five had an independent life. There was neither residual epilepsy requiring long term treatment nor secondary hydrocephalus in our patients.

The abrupt rupture of a cerebral AVM is infrequent in childhood. In most series, children represent less than 20% of the population.¹ Except in infancy where Gallen vein malformations and large pial AVMs resulting in congestive heart failure are predominant, hemorrhagic stroke is the primary manifestation of AVMs in 50 to 85% of the cases.⁷⁻⁹ Some cases have been reported after a trivial head trauma,¹⁰ but, most of the time, no predisposing events could be found. In 15% to 30% of the cases, seizures are the first clinical manifestations.^{11,12} A relationship between the type of initial presentation and the size of the AVM has been described, the small AVMs manifesting themselves by strokes and the large ones by seizure.¹ Deeply comatose children could represent 50% of the cases in the particular entity of cerebellar hemorrhages.¹³ The relative incidence in other locations could be estimated from 10% to 38% of the cases.^{2,5,7,14} It can be suspected that this incidence could be underestimated since undiagnosed or inadequately emergently treated acute hemorrhagic strokes could be a cause of out-of-hospital sudden deaths in previously healthy children.

Tracheal intubation and mechanical ventilation are the first interventions in comatose children. Moderate hypocapnia (PaCO_2 : 30-35 mmHg) can efficiently reduce raised ICP, but the risks of dramatically reducing the cerebral blood flow in ischemic areas with more profound hyperventilation has to be kept in mind.¹⁵ In the presence of life-threatening brain herniation, rapid infusion of mannitol 20% is effective in lowering raised ICP.¹⁶ The risk of increasing the volume of the intracranial hematoma, by decreasing the volume of the surrounding undamaged brain, is more theoretical than clinically relevant. When evidence of brain herniation is present, mannitol could be used as a life-saving therapy before craniotomy. Hypertonic saline solutions have the advantage of lowering raised ICP in the same range as mannitol with an associated effect of low volume resuscitation that can be mostly useful when hemodynamic instability is present.^{17,18} Direct ventricular puncture is another means of decreasing raised intracranial pressure that could be performed rapidly at the bedside. However, acute drainage resulting in ventricular collapse could result in a rapidly growing intraparenchymal hemorrhage and brain herniation.

In life-threatening situations, contrast-enhanced CT scan is the most easy and quickest examination to perform. It allows detection of intraparenchymal hematoma and intraventricular hemorrhage and

processes the degree of brain herniation. Supratentorial hematoma represent 70% to 80% of the cases^{5,8} and among them, 66% are hemispheric essentially in the parietal area. Deep-seated lesions develop in the basal ganglia, thalamus and corpus callosum. When the AVM is superficial, its rupture results in a parenchymal hematoma associated with subarachnoid hemorrhage. In deeply located lesions, massive intraventricular bleeding is more frequent and is an important factor in the increased mortality. Cerebellar hemorrhages are described as devastating conditions resulting in dramatic presentation with acute brainstem compression in 50% of the cases.¹³ We did not find a worse evolution in infratentorial localisation. If CT scan is sufficient to guide surgical decompression, it is insufficient to relate a hemorrhagic stroke to the rupture of an AVM. Obviously, it cannot allow a complete scheduled surgical resection without more precise preoperative identification of all the feeding arteries. MRI with three dimensional reconstruction and fast sequences is a very sensitive examination for the diagnosis of AVM, especially in angiographically occult AVM (AOAVM) that could be responsible for 10% of the hemorrhagic strokes so-called of unknown origin.⁵ The limited availability, the length of the examination, and the difficulties encountered in the management of unstable children in the MRI environment limit the use of MRI explorations in emergency situations. Cerebral angiography is the standard for exploring AVM. It could be falsely negative in as much as 10% of cases.⁵ Emergency angiography in children with intracranial hematoma carries a high risk of neurological and cardiovascular complications and of immediate rebleeding.¹⁴ Emergency endovascular treatment is not advisable in these cases, because it will not remove the brain compression resulting from the hematoma, which is the main life-threatening problem. Moreover, bleeding induces modification of the angioarchitecture of the AVM and reduces cerebral blood flow within the malformation resulting in difficult selective catheterization. As others, we decided to exclude preoperative angiography from the protocol of examinations that could be performed in deeply comatose children with evidence of brain herniation.^{5,19} Angiography can be proposed on a delayed emergency basis, and endovascular treatment could be applied to residual AVM. If a complete cure is obtained with embolization alone in less than 10% of the cases, a staged embolization is certainly useful associated with surgical resection for managing large AVM.^{20,21} This kind of management was successfully used in five of the children in this series.

The need for prompt evacuation of the hematoma and/or drainage of massive intraventricular thrombus

is not questionable.^{5,8,10,11} The same surgical management has been proposed in adults with profound neurological deterioration resulting from hemorrhagic stroke with mass effect.¹⁹ Such high-risk procedures have unpredictable blood loss and potential risks of intractable brain herniation through the dural incision. However, our 15% surgical mortality compares favourably, although the severity of the initial presentation was higher, with the 48% in children treated conservatively.¹² There is controversy concerning the need for complete excision of the AVM at the first operation. On one hand, complete excision, whenever possible, will prevent the risk of early rebleeding and result in neurologically normal children in 53% of the cases.⁵ On the other hand, it carries a high risk of increased blood loss and difficulty in controlling feeding pedicles that have not been clearly identified by precise preoperative neuroradiological evaluation. On the top of that, it also carries a higher risk of permanent neurological deficit when the hematoma lies in a functionally eloquent area. The rules that can be proposed for emergency management are to attempt excision of the AVM only when the hematoma is superficial and situated far away from an eloquent area. This is particularly indicated when the preoperative contrast enhanced CT scan showed the AVM. In all the other cases, a simple excision of the hematoma must be performed associated with the insertion of an external ventricular drainage when a ventricular hemorrhage is present.¹⁹

In the postoperative period, continuous measurement of ICP and CPP is of critical importance since the main risks in the postoperative period are the development of cerebral ischemia or, on the contrary, of massive cerebral swelling after brain decompression. Deep sedation prevents acute raised ICP related to stimulation, decreases cerebral metabolism,^{19,22} but precludes precise clinical examination. Instead of being based on a uniform protocol, critical care management should be adapted to the dynamic requirements of each patient.²³ The critical threshold for cerebral perfusion pressure below which hypoxia occurs is about 50 mmHg. The goal of a minimal CPP can be achieved by adequate volume expansion under central venous pressure and plasma osmolality control, and liberal use of vasoactive drugs such as dopamine.^{22,24} Massive intraventricular thrombosis could result in abrupt external drainage occlusion and acutely raised ICP. When classical measures have failed, direct intraventricular thrombolysis with recombinant tissue plasminogen activator is a safe and life saving method to restore shunt patency. It has the associated advantage of acting against cerebral vasospasm related to subarachnoidal hemorrhage.²⁵ In

the case of raised ICP related to postoperative brain edema, osmotherapy with mannitol 20% or hypertonic saline infusions are effective if the goal of a moderately increased plasma osmolality in the range of 300 to 310 mosmol·l⁻¹ can be achieved.²⁶ Rapid development of massive brain swelling after surgical decompression results in cerebral vasodilatation and acutely raised ICP.²¹ In the case of intractable raised ICP, severe hyperventilation should not be used until cerebral ischemia has been ruled out. Continuous jugular bulb oximetry is useful to separate global cerebral ischemia from hyperemia related to cerebral swelling.²⁷

Symptomatic cerebral vasospasm is a well-known complication of subarachnoidal hemorrhage that may occur in 46% of cases after aneurysmal subarachnoidal hemorrhage.²⁸ The incidence of vasospasm after pediatric AVM rupture is unknown. In the postoperative period, transcranial Doppler sonography measuring middle cerebral artery blood flow velocity can be performed easily. It allows diagnosis of persistent arterial spasm especially when concomitant measurement of the internal carotid artery flow velocity can be performed.²⁹ Nimodipine has proved its efficiency in preventing cerebral vasospasm related to subarachnoidal hemorrhage provided that normal circulating blood volume could be preserved.³⁰ With global ischemia unrelated to documented vasospasm, high-dose barbiturate coma has been used liberally by some authors.³³ The side effects, hemodynamic instability and increased rate of pulmonary complications, and the risk of increasing cerebral ischemia with vasocerebral vasoconstriction have to be kept in mind.³¹ It should be used as the last resort when other therapy has failed to control ICP.³²

Despite aggressive medical and surgical emergency management, the mortality remains high, but our mortality rate compares favourably with the 83% previously reported in children with Botterell V grade hemorrhage.⁵ This difference may be explained by a shorter delay between abrupt neurological deterioration and surgical decompression in our patients. More than the severity of the initial presentation or the precise location of the hematoma, the absence of response to initial critical care and the presence of a massive intraventricular hemorrhage seem to be factors responsible for the increased mortality.⁴ The trend toward increased mortality in cerebellar hemorrhage as compared to hemispheric hematoma⁵ was not noted in the present series. In survivors the reported functional outcome is good with minimal neurological sequels contrasting with the severity of the initial presentation.⁵ These good functional results are certainly an important argument for providing prompt and

aggressive critical care management whatever the initial presentation is.

It can be concluded that acute rupture of AVM resulting in deep coma still carries a high mortality. Prompt recognition of the diagnosis, early resuscitation, and adequate triage insure immediate survival. Even in moribund children, aggressive medical therapy could be efficient. There is no place, in this immediately life-threatening situation, for preoperative angiography that will not treat brain herniation. CT scan is the minimal radiological exploration before emergency surgical brain decompression. In survivors, aggressive emergency management and precise post-operative control of cerebral perfusion pressure, may result in a good functional outcome. Delayed angiography with graded embolization whenever possible and scheduled surgical complete excision or radiosurgery will complete this emergency treatment.

References

- 1 Mori K, Murata T, Hashimoto N, Handa H. Clinical analysis of arteriovenous malformations in children. *Child's Brain* 1980; 6: 13–25.
- 2 Fong D, Chan S. Arteriovenous malformations in children. *Child's Nerv Syst* 1988; 4: 199–203.
- 3 Eiras J, Gómez-Perún J, Carcavilla LI, Alberdi J. Surgical experience of arteriovenous malformations in children. *Child's Nerv Syst* 1987; 3: 156–60.
- 4 Takeshita M, Kagawa M, Izawa M, Kitamura K. Hemorrhagic stroke in infancy, childhood, and adolescence. *Surg Neurol* 1986; 26: 496–500.
- 5 Humphreys RP, Hoffman HJ, Drake JM, Rutka JT. Choices in the 1990's for the management of pediatric cerebral arteriovenous malformations. *Pediatr Neurosurg* 1996; 25: 277–85.
- 6 Jennett B, Bond M. Assessment of outcome after severe brain damage. *Lancet* 1975; 1: 480–4.
- 7 Lasjaunias P, Hui F, Zerah M, *et al.* Cerebral arteriovenous malformations in children. Management of 179 cases and review of the literature. *Child's Nerv Syst* 1995; 11: 66–79.
- 8 Celli P, Ferrante L, Palma L, Cavedon G. Cerebral arteriovenous malformations in children. Clinical features and outcome of treatment in children and in adults. *Surg Neurol* 1984; 22: 43–9.
- 9 Hladky J-P, Lejeune J-P, Blond S, Pruvo J-P, Dhellemmes P. Cerebral arteriovenous malformations in children: report on 62 cases. *Child's Nerv Syst* 1994; 10: 328–33.
- 10 Nishi T, Matsukado Y, Marubayashi T, Masumitsu T, Yamamoto H. Ruptures of arteriovenous malformations in children associated with trivial head trauma. *Surg Neurol* 1987; 28: 451–7.
- 11 Gerosa MA, Cappellotto P, Licata C, Iraci G, Pardatscher K, Fiore DL. Cerebral arteriovenous malformations in children. (56 cases). *Child's Brain* 1981; 8: 356–71.
- 12 Mazza C, Pasqualin A, Scienza R, Bazzan A, Da Pian R. Intracranial arteriovenous malformations in the pediatric age: experience with 24 cases. *Child's Brain* 1983; 10: 369–80.
- 13 Chadduck WM, Duong DH, Kast JM, Donahue DJ. Pediatric cerebellar hemorrhages. *Child's Nerv Syst* 1995; 11: 579–83.
- 14 Millar C, Bissonnette B, Humphreys RP. Cerebral arteriovenous malformations in children. *Can J Anaesth* 1994; 41: 321–31.
- 15 Skippen P, Secar M, Poskitt K, *et al.* Effect of hyperventilation on regional cerebral blood flow in head-injured children. *Crit Care Med* 1997; 25: 1402–7.
- 16 Mendelow AD, Teasdale GM, Russell T, Flood J, Patterson J, Murray GD. Effect of mannitol on cerebral blood flow and cerebral perfusion pressure in human head injury. *J Neurosurg* 1985; 63: 43–8.
- 17 Fisher B, Thomas D, Peterson B. Hypertonic saline lowers raised ICP in children after head trauma. *J Neurosurg Anesth* 1992; 4: 4–10.
- 18 Freshman SP, Battistella FD, Matteucci M, Wisner DH. Hypertonic saline (7.5%) versus mannitol: a comparison for treatment of acute head injuries. *J Trauma* 1993; 35: 344–8.
- 19 Jafar JJ, Rezaei AR. Acute surgical management of intracranial arteriovenous malformations. *Neurosurgery* 1994; 34: 8–13.
- 20 Fizzel RT, Fisher WS III. Cure, morbidity, and mortality associated with embolization of brain arteriovenous malformations: a review of 1246 patients in 32 series over a 35-year period. *Neurosurgery* 1995; 37: 1031–40.
- 21 Menovsky T, Van Overbeek JJ. Cerebral arteriovenous malformations in childhood: state of the art with special reference to treatment. *Eur J Pediatr* 1997; 156: 741–6.
- 22 Meyer P, Legros C, Orliaguet G. Critical care management of neurotrauma in children: new trends and perspectives. *Child's Nerv Syst* 1999; 15: 732–9.
- 23 Rosner MJ, Rosner SD, Johnson AH. Cerebral perfusion pressure: management protocol and clinical results. *J Neurosurg* 1995; 83: 949–62.
- 24 Pfenninger J. Neurological intensive care in children. *Intensive Care Med* 1993; 19: 243–50.
- 25 Öhman J, Servo A, Heiskanen O. Effects of intrathecal fibrinolytic therapy on clot lysis and vasospasm in patients with aneurysmal subarachnoid hemorrhage. *J Neurosurg* 1991; 75: 197–201.
- 26 Simma B, Burger R, Falk M, Sacher P, Fanconi S. A prospective randomized and controlled study of fluid

- management in children with severe head injury: lactated Ringer's solution versus hypertonic saline. *Crit Care Med* 1998; 26: 1265–70.
- 27 *Von Helden A, Schneider GH, Unterberg A, Lanksch WR.* Monitoring of jugular venous oxygen saturation in comatose patient with subarachnoid haemorrhage and intracerebral haematomas. *Acta Neurochir* 1993; 59(Suppl): 102–6.
- 28 *Solenski NJ, Haley EC Jr, Kassell NF, et al.* Medical complications of aneurysmal subarachnoid hemorrhage: a report of the multicenter, cooperative aneurysm study. *Crit Care Med* 1995; 23: 1007–17.
- 29 *Lindegaard KF, Nornes H, Bakke SJ, Sorteberg W, Nakstad P.* Cerebral vasospasm diagnosis by the means of angiography and blood velocity measurements. *Acta Neurochir* 1989; 100: 12–22.
- 30 *Auer LM.* Acute operation and preventive nimodipine improve outcome in patients with ruptured cerebral aneurysms. *Neurosurgery* 1984; 15: 57–66.
- 31 *Cruz J.* Adverse effects of pentobarbital on cerebral venous oxygenation of comatose patients with acute traumatic brain swelling: relationship to outcome. *J Neurosurg* 1996; 85: 758–61.
- 32 *Louis PT, Goddard-Finegold J, Fishman MA, Griggs JR, Stein F, Laurent JP.* Barbiturate and hyperventilation during intracranial hypertension. *Crit Care Med* 1993; 21: 1200–6.