Review Articles

Cerebral arteriovenous malformations in children

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The treatment of cerebral arteriovenous malformations (AVM) or vascular anomalies are challenging neurosurgical procedures for an anaesthetist. Large AVMs are uncommon in children. Only 18% of AVMs become symptomatic before the age of 15 yr. This series reviews the experience at this institution during the period of 1982 to 1992. The symptoms at the time of presentation are varied and include haemorrhage (50%), seizures and hydrocephalus (36%) or congestive cardiac failure (18%). Symptoms of congestive heart failure predominate in the newborn whilst neurological symptoms, such as stroke, seizures or hydrocephalus occur more commonly in infants and older children. Approximately one third of AVMs in childhood present acutely. Radiological investigations,. e.g., CT scan, MRI and cerebral angiography are essential to identify the precise location of the lesion. Therapeutic intervention in the acute presentation may involve craniotomy for evacuation of haematoma and treatment of increased intracranial pressure (ICP). Control of seizures and congestive heart failure may take priority and allow time to plan the elective procedures of embolization and surgical excision of the AVM. Operative intervention is hazardous and peroperative complications can be expected in more than 50% of patients. The morbidity and mortality associated with cerebral AVM are high, especially in infants who present in the neonatal period with congestive cardiac failure. The overall mortality in this series was 20%. Children presenting with intracranial arteriovenous malformations require a multidisciplinary approach. The successful management of anaesthesia either for embolization or surgical resection necessitates an understanding of the disciplines of paediatric and neuroanaesthesia. Special care and specific attention to detail may contribute to reduce the high morbidity and mortality encountered in these compromised children.

Le traitement neurochirurgical des malformations cérébrales artérioveineuses (MAV) représente pour l'anesthésie un défi majeur. Les grosses MAV sont rares chez l'enfant. Seulement 18% deviennent symptomatiques avant l'âge de 15 ans. D'après notre expérience de 1982 à 1992, les premières manifestations sont variables: hémorragie (50%), convulsions et hydrocéphalie (36%) ou défaillance cardiaque globale (18%). Les symptômes de la défaillance cardiaque globale prédominent chez le nouveau-né alors que les symptômes neurologiques, comme les convulsions et l'hydrocéphalie surviennent plus fréquemment chez le nourrison et l'enfant plus âgé. Environ un tiers des MAV de l'enfance se manifest subitement. Les études radiologiques comme la tomodensitométrie, l'imagerie par résonance magnétique et l'angiographie cérébrale sont essentielles pour l'identification précise du site de la lésion. Le traitement peut signifier une craniotomie pour l'évacuation d'un hématome et le traitement de l'hypertension intracrânienne. Le contrôle des convulsions et de la défaillance cardiaque est souvent prioritaire et permet de planifier des interventions programmées d'embolisation et d'excision chirurgicale de la MAV. L'intervention chirurgicale est dangereuse et on peut s'attendre à des complications peropératoires dans 50% des cas. La morbidité et la mortalité associées aux MAV sont élevées, spécialement en cas de défaillance cardiaque à la période néonatale. Dans notre série, la mortalité totale est de 20%. Ces cas doivent être traités par une équipe multidisciplinaire. Une conduite anesthésique efficace pour embolisation ou résection chirurgicale nécessite une bonne compréhension de l'anesthésie pédiatrique et neurochirurgicale. Des soins spéciaux et une attention particuliere aux détails peuvent contribuer à diminuer la morbidité et la mortalité élevées de ces enfants très malades.

Key words
ANAESTHESIA: paediatric, neuroanaesthesia;
SURGERY: paediatric, neurological arteriovenous malformation.

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Accepted for publication 27th November, 1993.

An arteriovenous malformation is a congenital vascular lesion that can arise anywhere in the body. Those occur-

ring in the brain may manifest in a multitude of symptoms and signs at any time in life. In the paediatric population, the commonest presentation is an intracerebral bleed, a seizure and hydrocephalus or, more rarely, congestive heart failure (CHF) in the neonatal period.

At the Hospital for Sick Children, Toronto, between 1982 to 1992, 154 procedures were undertaken in 56 paediatric patients with the diagnosis of cerebral arteriovenous malformation. The procedures can be grouped into four categories: (1) investigational – CT scan or cerebral angiography; (2) interventional radiology – embolization; (3) craniotomy; and (4) surgical procedures. This article will review the aetiology, presentation, and pathophysiology of AVM. It will also describe the anaesthetic techniques and discuss relevant issues surrounding the management of AVM in children.

Aetiology

The prevalence of the AVM lesion is difficult to assess. Autopsy studies have suggested that it is less than 1%; however, asymptomatic AVMs are discovered more frequently with the greater use of CT scanning.[2]

When all symptoms arising from an AVM are considered, the age at presentation is broad. The greatest number present between the ages of 20 to 45 yr, with a peak in the fourth decade, with a slight male preponderance. Why an AVM does not manifest itself until the ages of 20 to 45 yr remains a mystery. Only 18% of AVMs that become symptomatic do so before the age of 15 yr. Most remain quiescent until adulthood when they present with haemorrhage or seizures. The mortality from haemorrhage in a child is 25%. For all patients, adults and children, the annual mortality is estimated at between 1 and 2% with a 2.5 morbidity overall. Twenty years after diagnosis 10% of patients will have died, 22% will have mild deficits and 29% will be severely disabled.[3] The incidence of recurrent haemorrhage is estimated at 6% in the first year and 2% thereafter.[4]

They are usually congenital lesions which arise from the abnormal development of the arteriolar-capillary network which exists between the arterial and venous circulations within an organ. The stimulus, timing and early architectural deficiencies in the AVM are not clear. It is believed that the malformations occur before the embryo is 44-mm in length (the stage of definitive formation of the adult pattern of arterial wall structure).[1] Large cerebral AVMs are uncommon and usually well circumscribed. However, many are in the process of development and can become extensive. With growth of the brain there is progressive distention of the veins which leads to the formation of multiple fistulae. Eventually, the entire venous system of the brain and skull may become involved. The veins appear to be arterialized.

Anatomically, AVMs may be classified into three coexistent and distinct types:[5] (1) a tangled mass of tortuous and greatly dilated veins which pulsate and contain bright arterial blood; (2) an enlarged artery which supplies the lesion; (3) an interposed bed of dilated capillaries through which numerous arteriovenous communications are formed. In most cases the AVM is supratentorial and usually within the cerebral hemispheres. Only 24% are in the cerebellum and brain stem.[6] About 50% are believed to increase in size with age. Several studies[7,8] reveal that the potential for growth of an AVM is greatest in children and recommend that the lesions be excised surgically, even if they are small and have not ruptured.[9]

Cerebral damage may result from the presence of an AVM in neonates[10] due to several mechanisms including: (1) a "steal" phenomenon in the overlying abnormally dilated leptomeningeal vessels;[11] (2) ischaemia or failure of perfusion from congestive heart failure;[11] (3) haemorrhagic infarction from thrombosis of the aneurysm of the great vein of Galen;[11] (4) cerebral atrophy due to compression of adjacent structures by the aneurysm;[11] (5) alteration of flow caused by surgery.[10]

Knudson and Alden[12] in their review of 156 cases of AVMs in infants under six months of age noted that with the central nervous system lesions there was a male:female ratio of 2:1, with an incidence of CHF – 67%, bruits – 38%, cyanosis – 28%, murmur – 37% and increasing head circumference – 25%. the lesions occurred primarily in the vein of Galen (64%), in the cerebrum involving either the parenchyma, meninges or both (7%) together with the superior sagittal sinus (6%). The presenting signs and symptoms of cerebral AVMs were primarily of cardiac origin in the newborn but neurological in the infant.

Although the anatomical classification of AVMs has received much attention, no system is used universally. However, certain features may help in the assessment of risk, operability, morbidity and mortality. Pasqualin *et al.*[13] (in their review of 248 patients aged 1 to 62 yr) identified several prognostic features of AVMs:

1 *Volume of the lesion* (as opposed to diameter). A volume >20 cm^3 is associated with a higher incidence of intraoperative problems and postoperative hyperaemic complications. There is also more likelihood of new permanent deficits and a fatal outcome. This risk is even higher if the lesion is >50 cm^3.
2 *Feeding vessels.* The presence of deep feeding vessels increase the incidence of intra- and postoperative complications, as well as the morbidity and mortality.
3 *Shunt flow.* An estimation of flow by transcranial Doppler (TCD) shows that if the mean flow velocity in the main feeder vessel is >120 cm · sec and the pul-

satility index (PI)* is < 0.5 increased intraoperative blood loss and postoperative haematoma are more likely i.e., a negative correlation between flow and prognosis after surgical removal.

4 *Deep drainage system.* An extensive deep drainage system is associated more often with a poorer prognosis.

Pathophysiology

The presentation of a congenital AVM in children is primarily associated with symptoms of intracranial haemorrhage. Seizures and hydrocephalus, or congestive heart failure are less common presentations.

Haemorrhage

Intracranial arterial aneurysms and AVMs are collectively, though not exclusively, the commonest causes of spontaneous subarachnoid or intraventricular haemorrhage (SAH/IVH). Arteriovenous malformations account for 42% of haemorrhagic strokes in children[14] and SAH is the presenting feature of AVM in 77% of patients.[15] The fate of a child with an unexcised AVM is worse than that of an adult as there is a greater risk of bleeding (32% in ten years), especially in children with a non-haemorrhagic onset. The risk of haemorrhage does not decrease as the child grows older. The incidence of bleeding from an AVM is about 1% per year, but increases to 60% per year for recurrent haemorrhage.[4]

Calcium deposition in the walls of the affected vessels may be the cause of SAH. Thromboses may occur and evidence of repeated haemorrhages is often found at the time of surgical excision.

Seizure and hydrocephalus

Seizures occur as the first symptom in 25% of children with an AVM. They may occur at any age, but it has been estimated that up to 50% of patients with an AVM will have had one or more seizures by the age of 30 yr.

In infants, AVMs may present with increased ICP from obstructive hydrocephalus but the cause of the hydrocephalus has not been determined. Apart from the mass of the obstructing galenic lesion, it may be related to the blood flow in the AVM (i.e., size of the malformation) or to changes in the absorption of cerebral spinal fluid which may cause an increased ICP.[3]

*The pulsatility index (PI) is a measure of cerebrovascular resistance obtained from the recording of cerebral blood flow velocity and to a certain extent to cerebral blood flow. It is calculated by subtracting the diastolic from the systolic peak flow velocity and dividing the difference by the mean peak flow velocity; i.e., PI = (vs − vd)/vm.

TABLE I Age and sex distribution of the 56 children with cerebral AVM for the period 1982–92

Age at presentation	*n*	%	*M:F*
Birth to 1 mo	7	12.5	6:1
1 to 12 mo	6	11	4:2
1 to 5 yr	10	18	8:2
6 to 10 yr	17	30	7:10
11 to 18 yr	16	28.5	9:7
Total	56		34:22

TABLE II Presenting symptoms and sex distribution of the 56 children with cerebral AVM for the period 1982–92

Diagnosis	*n*	%	*M:F*
Intracerebral haemorrhage	28	50	16:12
Seizures	5	9	3:2
Headache	2	3.5	0:2
Hydrocephalus	5	9	4:1
Congestive heart failure	10	18	8:2
Other	6	10.5	3:3
Total	56		34:22

Congestive heart failure

Congestive heart failure (CHF) is the presenting symptom in less than one percent of AVM in the general population. However, in children it may account for a higher proportion. In neonates, it is often the sole presenting symptom. The AVM provides low resistance pathways for blood to return rapidly to the heart. Congestive heart failure results from an inability of the right and left ventricles to cope with the increase in venous flow. In the neonate the gradual decrease in pulmonary vascular resistance in the early post-natal period further contributes to the problem. The requirement for a high cardiac output to supply both these low-pressure circulations induces heart failure with a reduction in output to the other organ systems. Multi-organ failure secondary to CHF will further complicate the anaesthetic management. In general the outlook for symptomatic neonates is poor.[16]

Clinical presentation

The charts of 56 children presenting to The Hospital for Sick Children, Toronto for treatment of cerebral AVM in the years 1982–92 were reviewed. The ages at presentation and diagnoses are described in the Tables I and II. Compared with a previous review from our institution,[17] there was a fall in the number of intracerebral haemorrhages (72% to 50%) but an increase in the number of children treated who had an initial diagnosis of congestive heart failure (4.5% to 18%). Anaesthesia was administered on 154 occasions to these 56 children. Complete records were available for 149 anaesthetics.

Investigational procedures
A diagnostic CT scan was performed in the majority of children with appropriate sedation, but few required anaesthesia. A cerebral angiogram was performed in 91%; 22 of the 51 children had two or more angiograms. All angiographic studies were performed under general anaesthesia, making it the most common reason for general anaesthesia in this group of patients (Table III).

Interventional procedures
These included interventional radiology with intravascular embolization and/or neurosurgical intervention such as craniotomy for excision of the vascular anomaly (Table III).

Other procedures
Ten children with intracerebral AVM had anaesthetics for 15 other surgical procedures, including insertion of VP shunts, burr holes, orthopaedic operations and a corneal graft.

Anaesthetic management

Angiography
Emergency angiography was required in 19 of the 56 children reviewed. It accounted for 21 of the 77 angiographic procedures performed. Increased ICP was present in 86%. Elective angiography was required for diagnostic and reassessment purposes in another 43 children who underwent 56 procedures. Many had had emergency angiography on admission to this hospital.

PRE-OPERATIVE PREPARATION
Particular attention should be given to the neurological status (including ICP), patency of the airway, fasting state, cardiovascular stability and any therapeutic measures already undertaken. Measurement of haemoglobin concentration, haemocrit, serum electrolytes and blood cross-matching are routine.

When angiography is elective most patients are neurologically stable, fully fasted and frequently receiving routine anticonvulsant medication. In this series, 66% had received an anticonvulsant either alone or in combination with dexamethazone. Those children who presented with cardiac failure were maintained on their digoxin, diuretics, dopamine or other inotropic drugs. In the emergency situation premedication is not usually administered, but drugs used to control ICP such as mannitol or dexamethazone, and anticonvulsants such as diazepam, phenobarbitone and phenytoin may have been given. In this survey 29% of the emergency group had received one or more of these drugs before induction of anaesthesia.

TABLE III Distribution of emergency and routine anaesthetics given for angiography, embolization, craniotomy, CT scan and other procedures in the 56 patients during the period 1982–92

	Angiography	*Embolization*	*Craniotomy*	*Other*	*CT*
Emergency	21	0	16	6	3
Routine	56	15	25	10	0
Total	77	15	41	16	3

INDUCTION AND TRACHEAL INTUBATION
Anaesthesia induction techniques are dictated by the urgency of the clinical presentation. Tracheal intubation was required before admission to hospital or in the Emergency Department before diagnostic procedures were performed in 38%. In the remainder, rapid-sequence induction was used and included pre-oxygenation, cricoid pressure, thiopentone, atropine and succinylcholine to facilitate tracheal intubation.

In most patients intravenous induction of anaesthesia is suitable, although an inhalational induction may be useful, either alone or to facilitate intravenous access, induction being completed with thiopentone. Other induction agents are available. Propofol is known to have similar effects to thiopentone in attenuating the cardiovascular and ICP changes to manipulation of the airway.[18] Although ketamine raises ICP by increasing cerebral blood flow and remains contra-indicated in neuroanaesthesia, its lack of depressant action on the cardiovascular system may be beneficial in certain instances, i.e., in the child with congestive heart failure.

Atropine, *iv*, is advised before the use of succinylcholine to prevent bradycardia. Before tracheal intubation, lidocaine may be given *iv* to suppress pressor reflexes. Hyperventilation by bag and mask may be required in the presence of increased ICP.

MONITORING
Routine monitoring includes stethoscope, blood pressure, temperature probe, pulse oximeter, end-tidal CO_2 monitor and an airway pressure monitor with low-pressure alarms. An intravenous cannula (with as wide a bore as feasible) and an arterial cannula for continuous monitoring of arterial blood pressure are essential.

MAINTENANCE OF ANAESTHESIA
Nitrous oxide in oxygen with a low concentration of inhalational agent appropriate to the patient's clinical condition is commonly used. In this series isoflurane (48%) or halothane (24%) predominated but fentanyl alone was also satisfactory (24%). A neuroleptic technique of droperidol and fentanyl is an alternative to inhalational anaesthesia.

Pancuronium and d-tubocurarine were the most frequently used neuromuscular blocking agents (60% of patients). Some may not require relaxants and this may reflect the severity of the initial cerebral insult. Mild hypocapnia is considered useful in the control of intracranial hypertension, both in the emergency situation and after the operation. In this series 62% of all angiographic procedures has documented hypocapnia, with either a $P_{ET}CO_2$ or a $PaCO_2 < 35$ mmHg.

FLUID BALANCE AND BLOOD LOSS

The initial control of increased ICP often involves the administration of osmotic diuretics and fluid restriction. The aim is to reduce the amount free water available for oedema formation in the brain. A careful balance must be kept between this cerebral dehydration therapy and the maintenance of an adequate circulatory volume to ensure that cerebral perfusion continues to a "tight" brain. Angiography may result in a blood loss from the puncture site but, more importantly, may constitute a considerable fluid load in the small or compromised infant, where repeated injections of radio-opaque dye are required for visualization of the lesion.

EMERGENCE AND POSTOPERATIVE CARE

Some children require emergency craniotomy before decompression of the lesion after diagnostic angiography. Others require transfer to an intensive care unit for elective lung ventilation and planned craniotomy, and a few may proceed to embolization. The remainder will require no further treatment. In most elective patients the trachea can be extubated uneventfully at the end of the procedure after reversal of neuromuscular blockade. Rapid return of full consciousness is desirable to allow appropriate patient assessment in the post-anaesthetic recovery unit (PAR) since neurological complications or vascular compromise may occur.

COMPLICATIONS

These may be immediate or delayed. In this review more were noted in the emergency group (38%) than in the elective group (17%). Complications are best treated symptomatically and may be secondary to the initial cerebral insult (Table IV).

TABLE IV Complications following 77 angiographic procedures in this series

Complication	*Elective*	*Emergency*
Headache	2	0
Vomiting	2	0
Respiratory distress (elective), Arrest (emergency)	2	1
Drowsiness	1	3
Cool foot	1	2
Seizure	1	1
Embolus to ophthalmic artery – spontaneous resolution	1	0
CVS instability	0	1
Total	10	8

Craniotomy

Emergency craniotomy may be required soon after admission to hospital. In most cases an initial CT scan or angiographic procedure has been performed. The primary indication for craniotomy is evacuation of an intracerebral haematoma and treatment of raised intracranial pressure. The group reviewed consisted of 14 children undergoing 16 emergency craniotomies (88% with raised ICP) and 25 elective craniotomies in 23 children (35% with raised ICP).

PREOPERATIVE PREPARATION

This is similar to that described for angiography. Sedative premedication is rarely required in infants or younger children, but may be indicated for older children and teenagers. Therefore, a short-acting benzodiazepine agent such as midazolam may be the proper choice. Anticonvulsant therapy is routine. A haemoglobin concentration, haematocrit, serum electrolytes estimations are helpful. Cross-matched packed red cells (25% of the patient's estimated blood volume) must be available.

INDUCTION AND TRACHEAL INTUBATION

Monitoring devices are applied before induction of anaesthesia. Intra-arterial pressure monitoring is essential but, in children, placement of an arterial catheter should follow induction of anaesthesia to ensure optimum conditions. In this series of craniotomies the blood pressure was monitored directly in 90% of cases. In an emergency, many patients will arrive in the operating room from the radiology suite with a tracheal tube *in situ* (66%). Rapid sequence induction of anaesthesia is appropriate for the remainder. In the elective patient the mode of induction of anaesthesia is more variable, thiopentone is common and where ICP is not elevated, an inhalational induction may be preferred followed by a neuromuscular blocking agent to facilitate tracheal intubation. The use of lidocaine in elective craniotomy with normal ICP is debatable. This series revealed that in elective cases seven of 25 patients (28%) received *iv* lidocaine prior to intubation, only one of whom had documented evidence of increased ICP. In the emergency situation 87% of patients had elevated ICP yet only three (19%) received *iv* lidocaine.

MAINTENANCE OF ANAESTHESIA

The choice of technique depends upon the clinical state

of the patient. Nitrous oxide in oxygen with an inhalational agent is common and, recently, isoflurane in low concentration has assumed preeminence.[19] In this series the majority received isoflurane and in two-thirds of patients systolic BP decreased by more than 20% from baseline values. This may account for the relative decline in the use of specific hypotensive drugs. Pharmacological techniques for induced hypotension were used in only two of 25 elective craniotomies. A further three patients received high concentrations of isoflurane to produce hypotension. Other options for anaesthesia include a neuroleptic technique with droperidol and fentanyl, or fentanyl, nitrous oxide in oxygen and hyperventilation alone.[20] Intense neuromuscular blockade with pancuronium, d-tubocurarine or an infusion of atracurium or vecuronium is recommended, monitored with a peripheral nerve stimulator. Hyperventilation, to an $PETCO_2$ or $PaCO_2$ between 25 and 30 mmHg, is recommended.

Close attention must be given to integrity of the airway and breathing circuit and their relationship to positioning of the head. Not only can disconnection prove disastrous, but partial airway obstruction may have a detrimental effect of ICP.[21] Furthermore, because of the major determinant of ICP changes is intracranial volume, venous drainage obstruction may have a detrimental effect on ICP. The risk of air embolus increases with the head-up and sitting positions. Detection of air embolus is most rapid with a precordial Doppler probe, but an oesophageal stethoscope is also useful.

Blood loss at craniotomy is variable so it is advisable to have at least one large bore *iv* cannula for the rapid infusion of fluids. A blood warmer and pressure infuser are recommended. In our review of 40 craniotomies blood loss was estimated by the method used by McLeod.[17] If blood volume is assumed to be constant:

Total Haemoglobin (Hb) lost = (pre-op Hb + total transfused Hb) − 1st day Hb.

Blood loss is expressed as a percentage of estimated blood volume (EBV):

Total Hb lost/pre-op Hb × 100 = % lost.

Information for blood loss calculation was available on 32 of the 40 craniotomies: seven lost <10%, thirteen lost 10–20%, ten lost 20–50% of EBV. In two instances the blood loss was more than 50% of their EBV (68% and 302% respectively). If these two patients are excluded, the average % of EBV los was 16 ± 12%.

EMERGENCE AND POSTOPERATIVE CARE

Most emergency patients will require a period of postoperative lung ventilation and close neurological observation. This is best performed in the intensive care unit, although a well equipped high dependency unit (HDU) or PAR may suffice. Cerebral oedema resulting from the lesion or secondary to surgical manipulation and retraction may require further therapeutic intervention. Thus, return to full consciousness may be delayed for several days. After elective craniotomy the trachea will be extubated in most patients at the end of the procedure. However, close observation for neurological deterioration is required and the most appropriate setting is an ICU, HDU, or PAR.

TABLE V Complications following 40 craniotomies for AVM

Complication	*Emergency*	*Elective*
Death	3	1
Respiratory arrest	1	1
Seizure	1	3
Massive haemorrhage		2
Drowsiness	1	2
CVS instability		3
Swollen eye		1
Persistently high ICP		2
Respiratory depression		1
Headache		1
Electrolyte abnormalities – 2° to CHF	1	
Total	7	17

COMPLICATIONS

These may result from the lesion itself or from the therapeutic intervention. In this series complications arose in seven of 15 emergency cases (47%). This high incidence may indicate the severity of the presenting symptoms rather than anaesthetic or surgical complications. Complications were recorded in 14 of 25 elective cases (56%) (Table V).

Embolization

Interventional neuroradiology techniques are designed to ameliorate or cure the disorder without the need for subsequent invasive surgery. A transfemoral approach to the cerebral circulation under general anaesthesia allows access via the cervical internal carotid or vertebral artery. Location of the lesion with angiographic screening allows accurate placement of detachable balloons, silk sutures, platinum microcoils, polyvinyl alcohol sponges and polymerizing agents.[22] Subsequent thrombosis around these foreign bodies obliterates the lesion.

Embolization is a useful adjunct or alternative to surgical treatment of AVMs.[23] It may be particularly useful in the younger infant with CHF.[24] In this series, 15 embolization procedures (nine children) were performed. Of these, five patients presented with CHF and were receiving aggressive therapy prior to their procedure. Only one had documented evidence of raised ICP.

Preoperative preparation
Embolization is an elective procedure. A complete medical history and physical examination, assessment of neurological status, haemotological and biochemical review should be available. The presence of CHF may complicate the management of these children. Pre-medication with an anticonvulsant or dexamethazone may be required.

Induction and tracheal intubation
Preparation as for an elective craniotomy is recommended. Monitoring and intravenous access are established before induction. In this series, in neonates and infants without CHF, anaesthesia was induced with thiopentone, followed by atropine and succinylcholine to facilitate tracheal intubation.

Maintenance of anaesthesia
The considerations are similar to those for craniotomy. Where CHF is present the use of an inhalational agent may be limited due to its cardiovascular depressant effects. A supplementary dose of fentanyl may be useful. In this series hyperventilation to a $P_{ET}CO_2$ or $PaCO_2$ of <35 mmHg was recorded in 60%, but the CO_2 tension was elevated deliberately in 27% to produce vasodilatation of the cerebral vessels which may aid the passage of invasive catheters and visualization of the vascular lesion.

Emergence and postoperative care
In most children tracheal extubation will be performed at the end of the procedure. Close observation of the puncture sites and of neurological function is recommended for a few hours. Where CHF is present the patient should be transferred to the ICU for continued observation and therapy.

Complications
These include fluid overload from the radio-opaque dyes, from the lesion itself and deterioration due to the lesion itself or from a protracted anaesthetic. There were 11 complications from 15 cases in this series. The high death rate (4/15) may reflect the fact that the earlier cases were all infants with CHF who were considered to have surgically inoperable lesions (Table VI).

Follow-up
Death occurred in 11 of 56 patients (20%). The M:F ratio was 6:5 and the presenting diagnoses were: cerebrovascular accident (CVA) – five patients, CHF – four patients and hydrocephalus – two patients. Longer term follow-up revealed an overall good or satisfactory outcome in the majority of patients as assessed by return to family care and their achievement within their capabilities subsequent to the physical insult suffered at the time of presentation.

TABLE VI Complications following 15 embolization procedures in nine children

Death	4
Intracerebral haemorrhage	2
Seizure	2
Failure to embolize	2
Cool foot	2
Drowsiness	1
Perforation of ventricle	1
Respiratory failure	2

Discussion

Cerebral arteriovenous malformations are congenital vascular lesions involving the arteriolar-capillary network within the brain. The prevalence rate of AVM in the general population is estimated at 1%, but only 18% of these AVMs present before the age of 15 yr. Common manifestations in the paediatric population include intracerebral bleed (50%) or seizures and hydrocephalus (36%). Congestive heart failure in the neonatal period is a rarer occurrence (18% in this series). The mortality from haemorrhage in children is 25%.

No single system of classification is used universally, however, certain features of an AVM may assist in assessment of risk, operability, morbidity and mortality. These include the volume of the lesion, the origin of the feeding vessels, the shunt flow velocity and the presence of a deep drainage system.

General anaesthesia is required for cerebral angiography. It is performed in 91% of children presenting with an AVM. Increased intracranial pressure can be anticipated in 86% of patients. An emergency angiographic procedure is required in 27% of cases and will be repeated later for reassessment and follow-up purposes (Table III). Many patients proceed to craniotomy, either for evacuation of intracerebral haematoma and treatment of raised ICP or for excision of the lesion. Embolization is an elective neuroradiologic technique which aims to ameliorate or cure the disorder without recourse to surgery. It is not without risk but is particularly useful and highly indicated in the neonate with congestive heart failure.

Control of arterial blood pressure
Direct intra-arterial pressure monitoring is essential for the safe conduct of anaesthesia in these patients. Moreover, it provides access for blood gas analysis and continuous assessment of intravascular fluid status and cardiac contractility. Pharmacological reduction of systolic BP to facilitate surgical access, reduce intraoperative

blood loss and increase cerebral protection has been advocated.[25] In this series controlled hypotension was used in only five of 25 routine craniotomies (20% compared with 60% in the previous review from this institution[17]). Two patients received pharmacological agents (labetolol in a ten year old and pentolinium in a 13 yr old). In each instance the mean arterial pressure did not decrease below 60 mmHg. One explanation for the decline in the use of induced hypotension could be that blood loss is not less in patients who have had a period of induced hypotension for AVM resection.[17] Although this conclusion is not drawn from a randomised, controlled study, the prudent course is to decrease systolic BP to a level required for successful surgery. Isoflurane which produces moderate hypotension appears to be safe and sufficient for this purpose.[26]

The acceptable lower limit of blood pressure and the duration of controlled hypotension in children is not known. The normal cerebral circulation can regulate blood flow over a wide range of mean arterial pressures (MAP) whereas abnormal vasculature seems to lose this intrinsic mechanism. The break point for autoregulation in adults is a MAP of 50 mmHg.[27] This value has also applied to children without specific validation.[28] However, since cerebral perfusion pressure (CPP) is the difference between MAP and intracranial pressure (ICP), if ICP is elevated a similar increase in MAP is required to maintain an adequate CPP. Swedlow[29] proposed that induced hypotension should be avoided in the brain-injured child, since those with an increased systemic pressure may be physiologically adapting to maintain an adequate CPP in the face of an elevated ICP. Therefore, the use of a lower MAP could be detrimental and eventually lead to ischaemia.

Blood loss and fluid balance

In young children and infants undergoing neurosurgical procedures, blood loss can be difficult to assess from swabs, head drapes and suction bottles. Haemorrhage may be insidious or abrupt. Infiltration of the scalp with bupivacaine 0.125% and epinephrine 1:200,000 has been demonstrated to prevent subsequent haemodynamic responses to scalp incision and reflection. Furthermore, it minimizes the blood loss during flap reflection.[30] Induced hypotension has been advocated[31] for cerebral protective effects and to minimize the peroperative losses. This series has revealed that induced hypotension is used infrequently.

It is common practice to restrict *iv* maintenance fluids to two-thirds of the daily requirement to reduce the intake of free water which may exacerbate cerebral oedema. The use of osmotic diuretics to reduce brain mass and lower ICP will further complicate the assessment of fluid requirement especially in the presence of on-going blood loss.

The management of massive blood loss during craniotomy requires rapid restoration of circulating blood volume, oxygen carrying capacity, haemostatic capability, colloid osmotic pressure and biochemical balance. Fluid replacement with crystalloid and/or colloid continues until losses reach 40% of a blood volume when replacement of red cells is required.[32] In this series the average blood loss was 16% of a blood volume. Crystalloid and colloid (5% albumin) solutions were used as replacement. Of the 30 children who lost $<50\%$ of a blood volume half did not receive blood peroperatively.

If bleeding continues blood should be sent to the laboratory periodically for haemoglobin, haemocrit, platelet count, clotting screen and serum electrolyte assay. These tests will assist in deciding when fresh frozen plasma or platelet transfusions are required. Fresh frozen plasma should be restricted to controlling defined defects in the coagulation cascade, e.g., prolongation of the prothrombin time and activated clotting time by 1.5 times normal.[33] Functionally, a platelet count $>50 \times 10^9 \cdot L^{-1}$ is required for haemostasis. Transfusions of one unit of platelet concentrate per 10 kg will usually suffice to raise by $50 \times 10^9 \cdot L^{-1}$ the intravascular platelet count which would be enough for microvascular bleeding, in the absence of diffuse intravascular coagulation.[32]

Biochemical disturbances of calcium homeostasis are a theoretical concern, but rarely a clinical problem.[34] Hyperkalaemia is usually transient and less important, but the combination of hypocalcaemia and hyperkalaemia, exacerbated by hypothermia, may cause cardiac irregularities.[35]

For craniotomy the availability of two to four units of cross-matched packed red cells, and the facility for rapid transfusion of warmed blood products is advisable.

Cerebral protection

This is the "prevention or amelioration of neuronal damage evidenced by abnormalities in cerebral metabolism, histopathology or neurologic function occurring after a hypoxic or an ischaemic event."[36] Therapy is directed at both the prevention of ischaemic cerebral pathology and the resuscitation of tissue that has already sustained ischaemic damage. Neuroprotective agents act by reducing cerebral metabolic rate for oxygen ($CMRO_2$), improving oxygen delivery or by altering intracellular pathological processes.[37]

The routine use of cerebral protective manoeuvres has not been specifically addressed in the paediatric patient. In adults the use of agents to attenuate the cardiovascular responses to intubation have become routine. The assumption is that increases in ICP are attenuated by amel-

ioration of the hypertensive responses to laryngoscopy and intubation. The administration of lidocaine[38] *iv*, a second dose of induction agent (thiopentone[39] or propofol) or the administration of short-acting beta-blocking agents (esmolol[40]) have been advocated. Such practise has not been readily accepted for the paediatric population. However, the administration of an analgesic such as fentanyl or alfentanil[21] at induction may reduce the cardiovascular responses to laryngoscopy and intubation without the depressant effects of repeat dosing of induction agents.

Intravenous lidocaine in a neonate may cause severe cardiac dysrhythmias.[41] Splinter[42] using 1.5 $mg \cdot kg^{-1}$ of lidocaine *iv* in the 2–12 yr age group found that while there were no adverse effects it did not attenuate the pressor responses to laryngoscopy and intubation. This series of cases bears out the tendency not to use lidocaine in the paediatric population.

Hyperglycaemia may exacerbate the neurological consequences of a cerebral ischaemic event.[43] It is felt that glucose-containing solutions should be administered only for specific clinical indications.[44]

Hypothermia

Hypothermia reduces the cerebral metabolic rate for oxygen ($CMRO_2$). Profound hypothermia has been advocated[45] but the resulting complications (delayed drug clearance, slow reversal of muscle relaxants, decreased cardiac output, conduction abnormalities, attenuated hypoxic pulmonary vasoconstriction, altered platelet function, electrolyte abnormalities, and postoperative shivering)[46] must be balanced against the small gain in protection. Although mild hypothermia may be desirable even in paediatric neuroanaesthesia, one must remember that because cooling of small infants and children is rapid, measures to prevent excessive hypothermia should be ensured. The maximum benefit in cerebral protection occurs where core temperature decreases to 34°C.[47] If specific measures such as a warming blanket,[48] forced-air convective blanket, blood warmer for *iv* fluids, humidification of inspired gases,[49,50] warm operating room environment[48] and covers for exposed skin are used, temperature can be maintained and the risks of accidental severe hypothermia avoided.

Induced hypocapnia

Induced hypocapnia is commonly used in neurosurgical procedures to reduce cerebral blood flow and volume and consequently intracranial pressure. Moderate hypocapnia with a $P_{ET}CO_2$ or $PaCO_2$ of 25 to 35 mmHg is often used. Cerebral blood flow (CBF) varies linearly with $PaCO_2$ between 20 and 80 mmHg, and a 1 mmHg decrease in $PaCO_2$ results in a 4% decrease in CBF. The controversy of the "steal" phenomenon, which reflects the balance of changes in perfusion pressure associated with reduction in ICP and increases in vascular resistance from the hypocapnia, has not been resolved.[52] This series revealed that hypocapnia was utilized in two-thirds of cases for angiography and in a majority of craniotomies. In 27% of elective embolizations hypercapnia was permitted to enhance visualization and to facilitate the catheter migration within the cerebral vasculature.

Osmotic diuretics

Mannitol has been used to reduce brain mass and ICP. Slow infusion of 0.25–1.0 $G \cdot kg^{-1}$ of mannitol reduces ICP within 15 min without cardiovascular effects.[53] The decrease in ICP can be enhanced and prolonged by the use of furosemide.[54] Repeated *iv* boluses control ICP with little rebound rise in ICP. Caution is advised with mannitol due to its potential to cause renal dysfunction in the severely brain-injured patient.[55]

Mortality

In this series the mortality was 11 of 56 patients (20%). Half were secondary to CVA (10% overall) and this is comparable with other series.[56,57] In comparison with the previous series[17] a larger proportion of patients were neonates presenting congestive heart failure (4% of 18%). Previously these patients were considered to have inoperable lesions whereas there is now a tendency to treat them aggressively and successfully.[58] This may influence statistics related to morbidity and mortality from interventional procedures in the future.

Conclusion

Anaesthesia for children presenting with complications from an intracranial AVM requires an understanding of the combined disciplines of paediatric and neuroanaesthesia. The treatment of neonates with congestive heart failure secondary to the AVM may be optimized by use of non-surgical therapy, such as embolization techniques before definitive surgery. In the older child the prognosis following treatment of the AVM is good, but morbidity and mortality from the consequences of the initial insult remain significant.

Acknowledgement

The authors would like to thank Dr J.E.S. Relton for his assistance in the preparation of this manuscript.

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