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Clinical feature and outcome of pediatric cerebrovascular disease: a neurosurgical series

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Abstract To investigate the clinical features and surgical outcome of pediatric cerebrovascular disease (CVD), we retrospectively reviewed 280 children (up to 15 years of age) who underwent neurosurgical procedures for CVD between 1979 and 1998. Open surgery (*n*=448), endovascular procedures (*n*=22), and stereotactic radiosurgery (*n*=14) were the main neurosurgical procedures adopted. Clinical features and surgical outcomes were described according to the etiology of the CVD and the number of years of management. The mean duration of follow-up was 34 months. The mean age was 7.1 years, and the male-to-female ratio was 1.2:1. The most frequent CVD was moyamoya disease (62%). The surgical mortality related to CVD was 0.7%. Eighty-seven percent had Karnofsky Performance Scale scores of more than 70. We demonstrate increasing detection rates and improving outcomes in recent years. The clinical course in this study shows that recovery from CVD in children is good after neurosurgical procedures.

Keywords Cerebrovascular disease · Children · Surgical outcome

Introduction

The frequency of cerebrovascular disease (CVD) in children is about 2.5 per 100,000/year in the general population [21]. Recently, increased experience and improved radiologic techniques have led to a better understanding of pediatric CVD, which was once considered a rare disease [19]. A number of population-based registries [1, 20, 21] and medically oriented studies of pediatric CVD [4, 6, 8, 25] have been reported. However, surgical series of pediatric CVD have rarely been studied, and the majority of them have been limited to a specific category of CVD. CVD is responsible for about 10% of pediatric neurosurgical patients, many of which are treatable [16, 19]. Children tend to show better recovery after stroke than adults, because the immature brain is more plastic than the mature brain [6, 19]. Therefore, aggressive therapy often proves more rewarding in the pediatric field.

To investigate the clinical features and surgical outcome of pediatric CVD, we retrospectively reviewed 280 children (up to 15 years of age) who underwent neurosurgical procedures for CVD from 1979 to 1998. It is hoped that this study will provide the best available information for previously unanswered questions related to neurosurgical cases of pediatric CVD and should serve as a baseline for the assessment of the potential advantages of new treatment approaches.

Materials and methods

We retrospectively reviewed 280 children (up to 15 years of age) who underwent neurosurgical procedures for CVD at the Division of Pediatric Neurosurgery, Seoul National University Children's

Hospital between 1979 and 1998. All CVDs were defined by neuroradiological imaging and/or pathological examination. Children with hemorrhages related to tumor and trauma were excluded, as were those with extracranial vascular disorders. Four children with traumatic carotid cavernous fistula (CCF) and one with traumatic aneurysm were included. Medical records and neuroimaging data were reviewed.

Two hundred twenty-six patients underwent magnetic resonance imaging (MRI) and, prior to its availability, 45 patients were imaged by computed tomography (CT). In 7 patients, the diagnosis of posthemorrhagic hydrocephalus (PHH) was made by ultrasonography (US) only. Neuroradiological examination with angiography was performed in 246 children, being omitted in those for whom US, CT, and/or MRI were considered sufficient. Angiography was the sole neuroradiological investigation performed in 2 patients. Single photon emission computed tomography (SPECT) with ^{99m}Tc-hexamethylpropyleneaminoxime was carried out in 173 cases. Acetazolamide SPECT was performed in 113 patients with moyamoya disease (MMD). We evaluated the comprehensive cognitive function in 112 patients and intelligence quotients (I.Q.s) in 125 patients. The children's I.Q.s were determined according to the Korean Educational Development Institute-Wechsler Intelligence Scale for Children (KEDI-WISC). They were considered "normal" when either verbal or performance I.Q. was greater than 90. Recently, new diagnostic procedures including magnetic resonance angiography, diffusion/perfusion MRI, and positron emission tomography (PET) with [^{18}F]fluorodeoxyglucose (FDG) were performed in selected patients.

A total of 484 neurosurgical procedures were performed on these patients. Some children had more than one neurosurgical procedure. The three main neurosurgical procedures were open surgery (*n*=448), endovascular procedures (*n*=22), and stereotactic radiosurgery (*n*=14). During the same period, 3,511 neurosurgical management therapies were performed. Thus, CVD constitutes 14% of pediatric neurosurgical procedures. A variety of open surgical operations were performed according to the disease entity. In patients with MMD, surgery was usually performed in two stages, one side first and then the other by encephalo-duro-arterio-synangiosis (EDAS) and/or encephalo-myo-synangiosis (EMS) before October 1995. Subsequently most underwent staged operations by EDAS and/or bifrontal encephalo-galeo-synangiosis (EGS) [11]. Bifrontal EGS was performed during the first operation in some cases, and not during the second operation. Surgical resection of arteriovenous malformations (AVMs) was performed in 30 of 44 patients. All patients harboring PHH were treated with a ventriculoperitoneal (VP) shunt. In those patients with cavernous malformations, 13 patients had complete resection, while partial resection was performed in 3 children.

The endovascular procedures consisted in intravascular embolization (*n*=18) and detachable balloon occlusion (*n*=4). Intravascular embolization was carried out in 7 patients with AVM, 3 with arteriovenous fistula (AVF), and 1 with a vein of Galen aneurysm. Among these, 3 patients received multiple intravascular embolization therapy, 2 patients on three occasions and 1 patient on four. Detachable balloon occlusion was applied in 4 patients with CCF.

Linear accelerator radiosurgery (available from 1995) was performed in 2 patients with AVM. Gamma knife radiosurgery (available from 1997) was performed in 8 patients, 6 with AVMs and 2 with cavernous malformations. Both linear accelerator radiosurgery and gamma knife radiosurgery were used on 2 patients with AVM.

Clinical features and surgical outcome were assessed on the basis of the etiology of the CVD. To compare the incidence and outcome according to each CVD and the calendar years at the time of management, the 20-year study period was divided up as follows: 1979–1983, 1984–1988, 1989–1993, and 1994–1998. Patients were divided into four groups by age at the time of operation: less than 1 year old, 1–5 years, 6–10 years, and 11–15 years. Each patient's performance status was assessed using the Karnofsky Performance Scale (KPS) scoring method during follow-up.

The mean duration of follow-up was 34 months (range: 5 days to 155 months). Five patients were lost to follow-up.

Results

Overall 280 cases of pediatric CVD

There were in total 155 boys and 125 girls. The ages of the patients ranged from eighteen days to 15 years with a mean age of 7.1 years. Twenty-seven children were less than 1 year old, 74 children, between 1 and 5 years, 115 children, between 6 and 10 years, and 64 children, between 11 and 15 years. The distribution and demographics of CVD are shown in Table 1. The most frequent CVD was MMD (*n*=173), followed by AVM (*n*=44), PHH (*n*=19), and cavernous malformations (*n*=18). Temporal incidences of pediatric CVD are described in Table 2. The frequency of MMD increased rapidly after the first indirect bypass surgery in 1987. The incidence of AVM tended to be temporally constant, whereas that

Table 1 Number of cases, mean age and gender ratio of each pediatric cerebrovascular disease

Cerebrovascular disease entities	No. of cases (%)	Mean age (years)	Gender ratio (M:F)
Moyamoya disease	173 (62%)	7.1	1:1
Arteriovenous malformation	44 (16%)	10.0	1.6:1
Posthemorrhagic hydrocephalus	19 (7%)	0.3	1.7:1
Cavernous malformation	18 (6%)	8.8	1.3:1
Arteriovenous fistula	5	2.8	4:1
Aneurysm	5	9.2	2:3
Carotid cavernous fistula	4	7.0	3:1
Hemorrhagic disease of the newborn	4	0.1	3:1
Intracerebral hemorrhage	3	9.7	3:0
Middle cerebral artery stenosis	2	7.0	2:0
Venous infarction	1	12.0	1:0
Sturge-Weber syndrome	1	6.0	0:1
Vein of Galen aneurysm	1	1.4	1:0
Total	280 (100%)	7.1	1.2:1

Table 2 Temporal changes in the numbers of pediatric patients with cerebrovascular disease

Cerebrovascular disease entities	1979–1983	1984–1988	1989–1993	1994–1998	Total
Moyamoya disease	–	4	44	125	173
Arteriovenous malformation	–	12	17	15	44
Posthemorrhagic hydrocephalus	–	3	9	7	19
Cavernous malformation	–	2	7	9	18
Arteriovenous fistula	1	–	2	2	5
Aneurysm	–	3	–	2	5
Carotid cavernous fistula	–	1	2	1	4
Hemorrhagic disease of the newborn	3	1	–	–	4
Intracerebral hemorrhage	1	1	1	–	3
Middle cerebral artery stenosis	–	–	–	2	2
Venous infarction	–	–	–	1	1
Sturge-Weber syndrome	–	–	–	1	1
Vein of Galen aneurysm	–	–	–	1	1
Total	5	27	82	166	280

Table 3 Complications of each pediatric cerebrovascular disease

Cerebrovascular disease entities	Complications	No. of patients (%)
Moyamoya disease	Mortality	1 (0.6%)
	Infarction	17 (10%)
	Hematoma	3 (2%)
	Recurrent hemorrhage	1 (1%)
	Fluid collection at wound	5 (3%)
	Osteomyelitis of bone flap	1 (1%)
	Angiography morbidity	8 (5%)
Arteriovenous malformation	Hematoma	3 (7%)
	Recurrent hemorrhage	2 (5%)
	Infarction	1(2%)
	Radiation necrosis	1 (2%)
	Cyst formation	1 (2%)
	Seizure	1 (2%)
	Infection	1 (2%)
	Embolization morbidity	2 (5%)
Posthemorrhagic hydrocephalus	Malfunction	5 (26%)
	Infection	3 (16%)
	Seizure	3 (16%)
Cavernous malformation	Infarction	3 (17%)
	Recurrence	1 (6%)
	Seizure	1 (6%)
Arteriovenous fistula	Hematoma	1
	Seizure	1
Aneurysm	Infarction	1
Hemorrhagic disease of the newborn	Seizure	2
	Infarction	1
Intracerebral hemorrhage	Mortality	1
	Infarction	1
	Seizure	1

of cavernous malformation has increased rapidly since 1989. A variety of CVDs were detected between 1994 and 1998.

In Table 3, the complications of each type of CVD are described. The total mortality related to CVD during the 20 years of this study is 0.7% (2/280). A 2-year-old girl with MMD suffered from extensive cerebral infarction immediately after an EDAS operation. Preoperatively, the patient was neurologically free and MRI showed only moyamoya vessels without infarction. During the operation she showed severe brain swelling, and she died 5 days later in spite of intensive care. In the other patient with intracerebral hemorrhage (ICH), death followed poor clinical status prior to surgical intervention, and surgery offered no chance of clinical recovery. Among the 275 children in whom follow-up was available, 239 patients (87%) had a KPS score of more than 70 (Table 4). Patients with cavernous malformations, aneurysms, and AVMs had a good performance status; 100%, 100%, and 93% of these, respectively, had KPS scores of more than 70. In

Table 4 Cerebrovascular disease entities vs postoperative Karnofsky performance scores (*n*=275)

Cerebrovascular disease entities	KPS score > 70	KPS score ≤70	Total
Moyamoya disease	149 (87%)	22 (13%)	171
Arteriovenous malformation	39 (93%)	3 (7%)	42
Posthemorrhagic hydrocephalus	15 (79%)	4 (21%)	19
Cavernous malformation	18 (100%)	0 (0%)	18
Arteriovenous fistula	2	3	5
Aneurysm	5	0	5
Carotid cavernous fistula	4	0	4
Hemorrhagic disease of the newborn	1	2	3
Intracerebral hemorrhage	1	2	3
Middle cerebral artery stenosis	2	0	2
Venous infarction	1	0	1
Sturge-Weber syndrome	1	0	1
Vein of Galen aneurysm	1	0	1
Total	239 (87%)	36 (13%)	275 (100%)

Table 5 The number of years of management vs postoperative Karnofsky performance score (*n*=275)

Time period	KPS score >70	KPS score ≤70	Total
1979–1983	1 (0%)	3 (100%)	4
1984–1988	23 (85%)	4 (15%)	27
1989–1993	68 (86%)	11 (14%)	79
1994–1998	147 (89%)	18 (11%)	165
Total	239 (87%)	36 (13%)	275

contrast to this, only 33% of patients with hemorrhagic disease of the newborn (HDN), 33% of ICH, and 40% of arteriovenous fistula (AVF) had the same KPS scores (Table 4). The performance status of patients treated for CVDs tended to improve with time (Table 5). The distribution, complications, and KPS score of other CVDs are described in Tables 1, 3, and 4, respectively.

Moyamoya disease (Table 6)

The mean age of 173 MMD patients was 7.1 years (range: 7 months to 15 years). The male-to-female ratio was 1:1. The highest rate was observed in the age group from 6 to 10 years. The incidence of familial occurrence was 5%. Transient ischemic attack (TIA) with infarction was the most common clinical manifestation (53%). Eight patients (5%) had a history of intracranial hemorrhage. MRI showed evidence of infarction in 65% of patients and hemorrhage in 4%. Thirty-one percent of patients showed neither infarction nor hemorrhage. The most common Suzuki's angiographic stage [24] was grade 3 bilaterally (right 47%, left 43%). SPECT revealed perfusion defect in 30% of patients. Decreased perfusion was demonstrated in 66%. Eighty-four percent of patients with acetazolamide SPECT showed a decreased reserve. Of 107 patients who were evaluated for cognitive function, 93% had cognitive dysfunction. On the other hand, 63% of patients were mentally normal by intelligence evaluation (*n*=120).

Postoperative infarctions of variable size were the most common complication and occurred in 10% of patients (Table 3). Functional changes occurred in 13 of the 17 concerned. One hundred forty-nine patients (87%) had a KPS score of more than 70 (Table 4).

Arteriovenous malformation

The mean age of 44 AVM patients was 10.0 years, and the gender ratio (M:F) was 1.6:1. Thirty-one children presented with spontaneous ICH (intraparenchymal and/or intraventricular). Seven children had chronic seizure disturbance. The remaining manifestations were symptoms or signs of increased intracranial pressure and focal neurological deficits in 3 patients each. The AVMs were most often found in the cerebral hemisphere (*n*=29), followed in order of frequency by the thalamus/basal ganglia (*n*=4), cerebellum (*n*=4), ventricle (*n*=3), corpus callosum (*n*=3), and the brain stem (*n*=1). The size of AVMs ranged from 1 to 11 cm (mean: 3.1 cm) in diameter. Eight patients were identified as having co-existing intracranial aneurysms. Four patients had angiographically occult, histologically verified AVM. Postoperative hematoma (*n*=3) was the most common complication. Intravascular embolization-associated morbidity was noted in 2 patients, 1 case of each of catheter tip impingement and catheter tip fracture. In relation to linear accelerator radiosurgery, recurrent hemorrhage, radiation necrosis, and cyst cavity formation occurred in 1 case each. Thirty-nine patients (93%) had a KPS score of more than 70 (Table 4). Among 7 children with epilepsy, 6 were seizure free after open surgery; the other had a rare seizure after stereotactic radiosurgery.

Posthemorrhagic hydrocephalus

The mean age of 19 PHH patients was 4 months and the gender ratio (M:F) was 1.7:1. Twelve children had a his-

Table 6 Clinical features of 173 patients with moyamoya disease

Clinical features	No. of cases (%)		
Age	<1 year	3 (2%)	
	1–5 years	57 (33%)	
	6–10 years	85 (49%)	
	11–15 years	28 (16%)	
Family history	(+)	9 (5%)	
	(–)	164 (95%)	
Clinical manifestation	TIA only	59 (34%)	
	TIA with infarction	91 (53%)	
	Infarction	15 (9%)	
	Hemorrhage	8 (5%)	
MRI (*n*=173)	Infarction (–) and hemorrhage (–)	54 (31%)	
	Infarction (+)	113 (65%)	
	Hemorrhage (+)	6 (4%)	
		Right	Left
Angiographic stages[a] (*n*=172)	I	8 (5%)	11 (6%)
	II	24 (14%)	27 (16%)
	III	80 (47%)	74 (43%)
	IV	53 (31%)	53 (31%)
	V	5 (3%)	5 (3%)
	VI	0 (0%)	0 (0%)
	Normal	1 (1%)	1 (1%)
	Unknown	1 (1%)	1 (1%)
SPECT (*n*=167)	Perfusion defect	50 (30%)	
	Decreased perfusion	111 (66%)	
	Decreased reserve[b]	95 (84%)	
Comprehensive cerebral function (*n*=107)	Normal	8 (7%)	
	Abnormal	99 (93%)	
Intelligence quotient (*n*=120)	≥90	76 (63%)	
	<90	44 (37%)	

[a] The angiographic stages were evaluated with the criteria of Suzuki [24]
[b] Acetazolamide SPECT was performed in 113 patients

tory of premature delivery, detected in 2 cases by fetal sonography. Two had a hypoxic history. Three patients were born at full term with unremarkable perinatal histories. Five children experienced mechanical shunt malfunctions, and 3 suffered from infective complications. Seizures occurred in 3 patients during the follow-up period. Fifteen patients (79%) had a KPS score of more than 70 (Table 4).

Cavernous malformation

The mean age of 18 cavernous malformation patients was 8.8 years, and the gender ratio (M:F) was 1.3:1. The presenting symptoms were hemorrhage in 9 children, epilepsy in 4, hemorrhage and epilepsy in 4, and focal neurological deficits in 1 child. In this series, 16 cavernous malformations were located in the supratentorial space, with 1 case each in the cerebellum and the brain stem. The size of the cavernous malformations ranged from 0.8 to 6 cm (mean: 3.2 cm in diameter). A KPS score of more than 70 was achieved in all children. Among 8 children who had experienced seizure, all were free of seizure after neurosurgical procedures: 6 after surgical resection of the cavernous malformation and 2 after gamma knife radiosurgery. Six patients successfully discontinued antiepileptic drugs (AEDs); 5 after open surgery and 1 after radiosurgery.

Others

The mean age of 5 aneurysm patients was 9.2 years. There were 2 boys and 3 girls. Four of these patients with aneurysms presented with subarachnoid hemorrhage. A mass effect caused by giant aneurysm was found in 1 patient. Hunt-Hess grades were grade two in 4 cases and grade three in 1. There were 3 anterior cerebral-anterior communicating artery aneurysms, 1 internal carotid artery bifurcation, and 1 posterior cerebral artery aneurysm. Vasospasm occurred in only 1 patient with good clinical outcome. All these patients had a KPS score of more than 70.

The mean age of 5 AVF patients was 2.8 years, and there were 4 boys and 1 girl. No patients with open surgery (*n*=2) had a KPS score of more than 70, 1 of 3 patients with intravascular embolization also scored less than 70.

The mean age of 4 CCF patients was 7.0 years. There were 3 boys and 1 girl. All patients received DBO, and their clinical outcomes were excellent.

The mean age of 4 patients with HDNs was 1 month, and all were boys. All HDNs occurred after 1 month of age. The incidence of HDN has reduced in recent years. The surgical outcome in this group was not good. Only 1 of 3 patients who were followed up had KPS scores of more than 70.

The mean age of 3 children with ICHs was 9.7 years, and all were boys. The suspected etiology was cryptic AVM in all cases. The operation consisted of a clot evacuation in 2 and the placement of a ventriculostomy tube only, to remedy associated hydrocephalus in 1. The clinical outcome was poor in this group, and only 1 had KPS score of more than 70 after the neurosurgical procedure.

Discussion

As shown in this study, 14% of pediatric neurosurgical procedures are aimed at CVD. Therefore, pediatric CVD is no longer uncommon to neurosurgeons. Although ours is neither a population-based registry nor a medically oriented registry, we believe that the information yielded by this study will be of real value, because the clinical detail and surgical outcome of pediatric CVD are described. Most published series have classified pediatric CVD into ischemic and hemorrhagic stroke [1, 4, 6, 21, 25]. However, a variety of diseases may contribute to ischemic and hemorrhagic stroke. In addition, advanced neuroradiological imagings now make accurate diagnosis possible. Therefore, we assessed clinical features and surgical outcomes according to the etiology of the CVD and the number of years of management.

In pediatric CVD, it is known that the incidence rate of stroke is higher in boys than in girls [25]. In our study, boys also showed a higher incidence rate than girls. In our population there was a preponderance of children between 6 and 10 years of age. Owing to its unique pathogenesis, patients with HDN were detected at a young age. According to the classification by age of onset [23], all our patients were classed as late HDN. The mean age of patients with PHH, vein of Galen aneurysm, and AVF was less than 3 years. Early detection of these entities may be related to the combined hydrocephalus, which could be detected easily in these young children.

Contrary to the experience of western countries, MMD was found to be the major component of pediatric CVD in this study. Many MMD patients have visited our institute since the introduction of MRI. AVM has shown a relatively constant incidence rate over time, whereas that of cavernous malformation has increased rapidly in the recent years. These differences are almost certainly caused by the introduction of MRI. Improvements in diagnostic techniques have also led to the detection of diverse CVDs between 1994 and 1998.

Reported mortality rates have ranged from 11% to 37% [1, 4, 8, 10, 25]. The mortality rate of those with hemorrhage was considered to be higher than that for cerebral infarction [1, 4, 19, 21, 25]. During the 20 years of this study, the total mortality related to CVD was 0.7% (2/280), and 87% of patients were found to have a KPS score of more than 70. There are several possible explanations for this favorable result. First, the surgical outcome of pediatric CVD has changed dramatically in recent years as a result of new technologies such as microsurgery, endovascular procedures, and stereotactic radiosurgery. Secondly, the ability of the nervous system of a child to recover after surgery is often greater than that of an adult's, because the immature brain is more plastic than the mature brain [6, 19]. Thirdly, underlying vascular anomalies frequently seen in children are surgically treatable, whereas atheromatous vessels are commonly found in the aged [19, 20]. Therefore, the importance of surgical procedure may be greater in children than in adults. Finally, the findings of this study were probably biased by the fact that children with critical conditions might have been excluded, because this study was a surgery-oriented review.

MMD is characterized by a progressive occlusion of the internal carotid artery or its terminal branches, accompanied by the formation of extensive collateral vessels ("moyamoya" vessels) at the base of the brain. Characteristic clinical features of our study were no gender dominance (male-to-female ratio 1:1), frequent onset in the age group between 6 and 10 years (49%), the rate of familial cases around 5%, and the high incidence of symptomatic ischemia (95%). The most surprising finding that arose from the neuropsychological testing was that most children had some degree of cognitive dysfunction in spite of normal I.Q.s. This result demonstrates the importance of comprehensive neuropsychological testing and the revascularization procedure for ischemic ACA territory. In our institute, bifrontal EGS [11] has been combined with EDAS since 1995, to prevent progressive cognitive dysfunction and/or frequent paraparesis resulting from ACA ischemia. Although the clinical features of MMD are better known, MMD has long been a clinical entity of unknown etiology. The higher incidence of the disease in Asians and the familial occurrence suggest that MMD may have a genetic background [5]. Therefore, basic research on the pathogenesis is necessary.

Our observations of pediatric AVM are consistent with previous reports [2, 9]. There is a tendency to hemorrhage (71%), smaller lesions (mean; 3.1 cm diameter), and location in the posterior fossa (11%). Although open microsurgery provides the best chance of cure, endovascular embolization could be useful in managing AVM, alone or in combination with surgery or stereotactic radiosurgery [9, 13]. However, their relatively small size and tiny vessels often prevent easy cannulation for endovascular obliteration [9]. We experienced 2 cases of embolization-associated morbidity due to this unique diffi-

culty in pediatric endovascular techniques for brain AVMs. Stereotactic radiosurgery will be used increasingly in children to obliterate these small lesions, which are not associated with hemorrhage or are the residua of an operation [18]. Unfortunately, a number of radiosurgery-associated complications were observed, including rebleeding, radiation necrosis, and cyst cavity formation. Therefore, to clarify the role of radiosurgery in the treatment of AVMs in children further long-term study in a comprehensive series is needed.

In the month following the onset of PHH, the progression continues in 35% of patients and 5% eventually require the placement of a VP shunt [7]. Because these patients tend to be extremely small or medically unstable, they are often poor surgical candidates [7]. Additionally, surgical outcome is not good in these patients [7]. In this study, mechanical malfunction and infectious complications of patients with PHH were more frequent than in our previous reported series of patients with CSF shunting procedures: mechanical 26% vs 12% and infectious 16% vs 4% [14].

Before the availability of CT and MRI, the diagnosis of cavernous malformation was rarely made prior to surgery because it could not be demonstrated by angiography. Our experience suggests that open surgery is the best therapeutic modality for cerebral cavernous malformation, because of the high rate of success in controlling the clinical symptoms and the few complications [3]. Some authors consider that stereotactic radiosurgery is an effective strategy for seizure control and hemorrhage reduction [12], while others believe that stereotactic radiosurgery eliminates neither the risk of hemorrhage nor the lesion in the case of cavernous malformation [22]. Although the number of patients treated with radiosurgery was small in our series, all patients showed improvement in seizure control. On the other hand, complete obliteration of cavernous malformation is still not evident.

Intracranial aneurysms in childhood are extremely rare, particularly in the first decade of life. Only 1–2% of all aneurysms encountered are in patients under 18 years old [17]. A previously reported study on the characteristics of pediatric aneurysms [15] included the following findings: a male predominance in the neonatal and infant periods and a similar gender incidence in adolescence, a high frequency of terminal carotid bifurcation and of vertebrobasilar aneurysms, frequently encountered giant aneurysms, presentation with good clinical grades, and good outcome after surgery. Compared with earlier pediatric series, our study showed a female predominance and frequent anterior communicating artery aneurysms. It was similar to other studies in that the patients presented in good clinical grade and showed excellent recovery after operation. One patient who had a postoperative infarction also has a good quality of life.

In AVF and CCF, endovascular procedures are the main treatment strategies in our institution. They have led to favorable outcomes in all patients with CCF and were also used in 3 patients with AVF. Two of them had a KPS of more than 70. The poor outcome of AVF is probably related to the low ages of our patients and the associated brain anomalies. All patients with poor outcomes were less than 2 years old and presented with associated hydrocephalus.

As shown in this study, the incidence of HDN has been reduced by effective vitamin K replacement. Considering the high incidence of intracranial bleeding and poor surgical outcome of HDN, prevention by vitamin K replacement is a most effective measure.

Two of 3 patients with ICH had a poor clinical outcome. This might be due to poor clinical status prior to the operation or to inadequate control of the underlying cause of ICH.

Conclusion

This retrospective analysis of the treatment of 280 children with CVD revealed rates of detection and improving outcomes, which were related to more efficient diagnosis and treatment. The clinical course of this surgical series showed that the recovery of children after CVD tends to be good. Further study of the risk factors and basic research upon each type of CVD would allow the design of a more effective strategy for CVD management.

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References

1. Broderick J, Talbot GT, Prenger E, Leach A, Brott T (1993) Stroke in children within a major metropolitan area: the surprising importance of intracerebral hemorrhage. J Child Neurol 8:250–255
2. Celli P, Ferrante L, Palma L, Cavedon G (1984) Cerebral arteriovenous malformations in children. Clinical features and outcome of treatment in children and in adults. Surg Neurol 22:43–49
3. Di Rocco C, Iannelli A, Tamburrini G (1997) Surgical management of paediatric cerebral cavernomas. J Neurosurg Sci 41:343–347
4. Eeg-Olofsson O, Ringheim Y (1983) Stroke in children. Clinical characteristics and prognosis. Acta Paediatr Scand 72:391–395
5. Fukui M (1997) Current state of study on moyamoya disease in Japan. Surg Neurol 47:138–143

6. Giroud M, Lemesle M, Madinier G, Manceau E, Osseby GV, Dumas R (1997) Stroke in children under 16 years of age. Clinical and etiological difference with adults. Acta Neurol Scand 96:401–406
7. Hansen AR, Snyder EY (1998) Medical management of neonatal post-hemorrhagic hydrocephalus. Neurosurg Clin North Am 9:95–104
8. Higgins JJ, Kammerman LA, Fitz CR (1991) Predictors of survival and characteristics of childhood stroke. Neuropediatrics 22:190–193
9. Humphreys RP, Hoffman HJ, Drake JM, Rutka JT (1996) Choices in the 1990s for the management of pediatric cerebral arteriovenous malformations. Pediatr Neurosurg 25:277–285
10. Isler W (1984) Stroke in childhood and adolescence. Eur Neurol 23:421–424
11. Kinugasa K, Mandai S, Tokunaga K, Kamata I, Sugiu K, Handa A, Ohmoto T (1994) Ribbon enchephalo-duro-arterio-myo-synangiosis for moyamoya disease. Surg Neurol 41:455–461
12. Kondziolka D, Lunsford LD, Flickinger JC, Kestle JR (1995) Reduction of hemorrhage risk after stereotactic radiosurgery for cavernous malformations. J Neurosurg 83:825–831
13. Lasjaunias P, Hui F, Zerah M, Garcia-Monaco R, Malherbe V, Rodesch G, Tanaka A, Alvarez H (1995) Cerebral arteriovenous malformations in children. Management of 179 consecutive cases and review of the literature. Child's Nerv Syst 11:66–79
14. Lee JY, Wang KC, Cho BK (1995) Functioning periods and complications of 246 cerebrospinal fluid shunting procedures in 208 children. J Korean Med Sci 10:275–280
15. Meyer FB, Sundt TM Jr, Fode NC, Morgan MK, Forbes GS, Mellinger JF (1989) Cerebral aneurysms in childhood and adolescence. J Neurosurg 70:420–425
16. Mickle JP, Glasser RS (1996) Cerebrovascular disease in childhood. In: Youmans JR (ed) Neurological surgery. Saunders, Philadelphia, pp 1243–1263
17. Norris JS, Wallace MC (1998) Pediatric intracranial aneurysms. Neurosurg Clin North Am 9:557–563
18. Riva D, Pantaleoni C, Devoti M, Lindquist C, Steiner L, Giorgi C (1997) Radiosurgery for cerebral AVMs in children and adolescents: the neurobehavioral outcome. J Neurosurg 86:207–210
19. Roach ES, Garcia JC, McLean WT Jr (1984) Cerebrovascular disease in children. Am Fam Physician 30:215–227
20. Satoh S, Shirane R, Yoshimoto T (1991) Clinical survey of ischemic cerebrovascular disease in children in a district of Japan. Stroke 22:586–589
21. Schoenberg BS, Mellinger JF, Schoenberg DG (1978) Cerebrovascular disease in infants and children: a study of incidence, clinical features, and survival. Neurology 28:763–768
22. Seo Y, Fukuoka S, Takanashi M, Nakagawara J, Suematsu K, Nakamura J, Nagashima K (1995) Gamma knife surgery for angiographically occult vascular malformations. Stereotact Funct Neurosurg 64 Suppl 1:98–109
23. Sutor AH, Kries R von, Cornelissen EA, McNinch AW, Andrew M (1999) Vitamin K deficiency bleeding (VKDB) in infancy. ISTH Pediatric/Perinatal Subcommittee. International Society on Thrombosis and Haemostasis. Thromb Haemost 81:456–461
24. Suzuki J, Kodama N (1983) Moyamoya disease – a review. Stroke 14:104–109
25. Wang LH, Young C, Lin HC, Wang PJ, Lee WT, Shen YZ (1998) Strokes in children: a medical center-based study. Chung Hua Min Kuo Hsiao Erh Ko I Hsueh Hui St Chi 39:242–246