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Arterio-venous malformations in childhood: clinical presentation, results after operative treatment and long-term follow-up

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Abstract

In a series of 182 arterio-venous malformations (AVM) recently published [8] we found 33 children aged 1 to 16 years. In 28 cases, the admitting condition was hemorrhage. For 31 AVMs total excision was possible. We observed one postoperative death accounting for a mortality of 3%. All children but two were followed-up by personal examination. Two were not able to work due to severe neurological deficits and seven had not been able to get into their intended occupation.

Keywords: Arterio-venous malformation, children, hemorrhage, long-term follow-up.

1 Introduction

Numerous papers have been published on arterio-venous malformations during the last decades. Although AVMs are responsible for 30–50% of the hemorrhagic strokes in childhood, only few authors paid attention to the problems of this special age group [1, 5, 6, 10, 13]. Besides discussion of the natural history of AVMs and the question, which of them are likely to bleed and should be operated upon, no attempt has been made for a long-term follow-up after successful surgical excision of AVMs in childhood.

2 Patients

Between 1955 and 1985 33 children aged 1 to 16 years were operated upon at the Neurosurgical Department of the Johannes Gutenberg-University, Mainz, FRG. There were 18 boys and 15 girls. In 28 cases the admitting condition was cerebral hemorrhage. CT-scan revealed space occupying hematoma 23 of these cases, 5 showed only subarachnoid hemorrhage without clot formation. Of the cases that did not bleed, three presented with epileptic seizures, one showed a hemiparesis of the infantile type and one had an extracranial extension.

Angiography was done in all cases. Twelve of the AVMs with a maximum diameter up to 2 cm were classified "small", 14 as "medium-sized" (diameter 2–4 cm), seven as large (diameter more than 4 cm). Histological examination revealed 31 arterio-venous and two cavernous angiomas.

3 Operative treatment

In 31 AVMs total excision was possible. Two malformations seemed not to be excisable but had to be treated by ventriculo-peritoneal shunting due to hydrocephalus. We observed one postoperative death, accounting for a mortality of 3%. (Mortality in our complete series of 182 surgically removed angiomas was 10.3%.) One child with a large midline angioma of the basal ganglia, that had seemed not to be excisable, died one year after the ventriculo-peritoneal shunting procedure due to recurrent hemorrhage.

4 Results

All children but two have been followed up by personal examination in our clinic (seven from 1 to 5 years follwing surgery, 12 from 5 to 10 years, six from 10 to 20 years, four from 20 to 30 years). Twenty-one patients didn't show any or just min-

imal neurological deficit and were able to work when school was finished. Six of our patients were working although a greater neurological deficit, such as hemiparesis or aphasia, had remained. Two were not able to work due to severe neurological deficit. Twenty-two patients were able to realize their intended occupation after operation. Seven were not able to do so. Twenty-four patients reported no epileptic seizures more than one year after operation. Most of them were not on medication. Four patients had seizures controlled by anticonvulsive drugs. One child suffered from a severe epilepsy with marked personality changes.

We classified the results in four groups: "excellent" (1), "good" (2), "fair" (3), and "poor" (4).

Table I shows the importance of the site of an AVM for the final results. It is obvious that angiomas located in functionally important brain areas (parietal lobe, central region) show worse results after operation. However, it has to be kept in mind that bleeding in these cases led to a severe neurological deficit even before surgery. Table II shows the importance of the size of an angioma for the postoperative result. Although small and medium-sized angiomas have a smaller risk of postoperative deficits, it has to be emphasized that even large angiomas with many arterial feeders can be excised with little postoperative deficit. Compared to the adults in our complete series of 182 surgically removed AVMs, the postoperative deficits after removal of large AVMs and AVMs located in functionally important brain areas are markedly less.

Table I. Localization of angiomas and operative results.

Localization	Result/No. of cases					Total
	1	2	3	4	Died	
Frontal	5	1	–	–	–	6
Temporal	5	2	–	–	–	7
Parietal	–	3	–	1	1	5
Occipital	–	2	–	–	–	2
Sylvian fissure	1	–	–	–	–	1
Central region	1	3	3	–	–	7
Lateral ventricle	–	1	–	–	–	1
Corpus callosum, basal ganglia and midbrain	1	–	–	1	1	3
Cerebellum	–	1	–	–	–	1
Total	13	13	3	2	2	33

Table II. Size of angiomas and operative results.

Result	Diameter (cm)			Total
	2	2–4	>4	
1 Excellent	6	6	1	13
2 Good	4	6	3	13
3 Fair	1	1	1	3
4 Poor	1	–	1	2
Died	1	–	1	2
Total	13	13	7	33

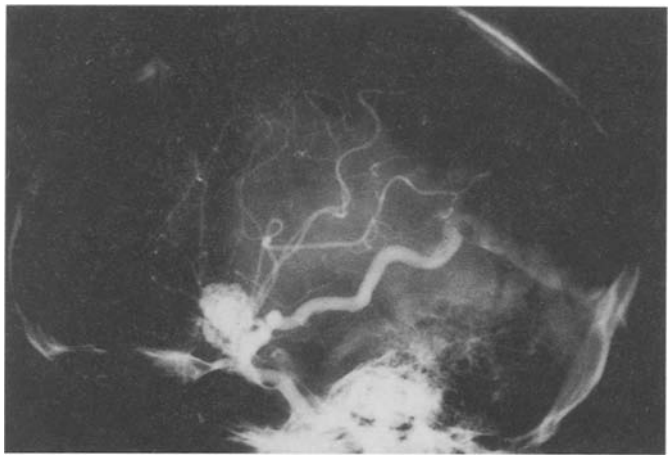
5 Case reports

Case 1: A 12 year-old girl very rapidly developed hemiplegia and became unconscious. Angiography revealed an AVM in the right Sylvian fissure arising from branches of the middle cerebral artery (Figures 1a, b). Craniotomy was performed eight weeks after bleeding and the AVM was removed completely, as could be demonstrated by postoperative angiography (Figures 1c, d). The patient was followed up to 15 years after operation. Meanwhile she has finished school and is working as a nurse without any neurological deficit.

Case 2: A boy aged 16 years had a subarachnoid hemorrhage with neck stiffness but no further neurological deficit. CT-scan did not demonstrate abnormalities but angiography revealed an AVM filled from the left pericallosal and the posterior cerebral artery with venous drainage to the vein of Galen (Figures 2a–c). Operation was performed two weeks after bleeding and the angioma could be successfully removed (Figures 2d, e). Five years after operation the young man had graduated from college and did not show any neurological deficit.

Case 3: While playing soccer, a 14 year-old boy sustained a massive intraventricular hemorrhage with clot formation and severe hemiparesis. Angiography revealed an arterio-venous malformation arising from the left choroid plexus (Figure 3a). Craniotomy was performed two weeks later and the angioma was removed. Postoperative angiographic figures were suspicious of demonstrating a small rest of AVM still filled (Figure 3b). The patient recovered well with only a minimal residual hemiparesis and about three epileptic seizures a year. He is on anticonvulsive drugs but is fully able to work.

Case 4: A girl aged 10 years had a subarachnoid hemorrhage and gradually became unconscious. She was admitted in a deep comatous state; CT-scans revealed an intracerebellar clot-formation causing obstructive hydrocephalus. Ventriculo-peritoneal shunting procedure was performed and the girl recovered. Angiography demonstrated an AVM of the antero-medial part of the right cerebellar hemisphere supplied by the anterior-superior cerebellar artery. It was possible to remove the



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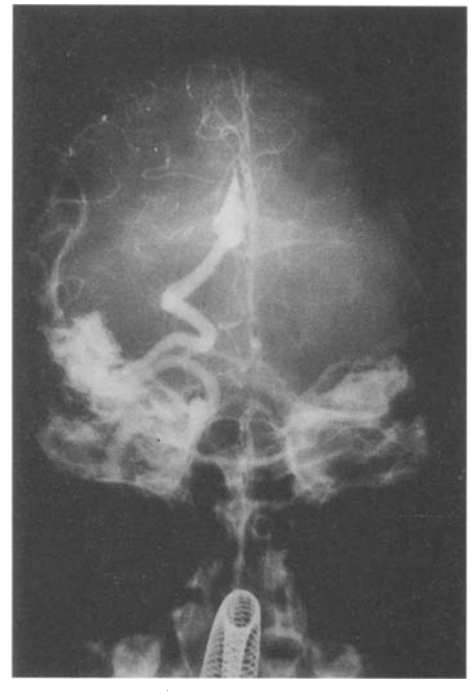

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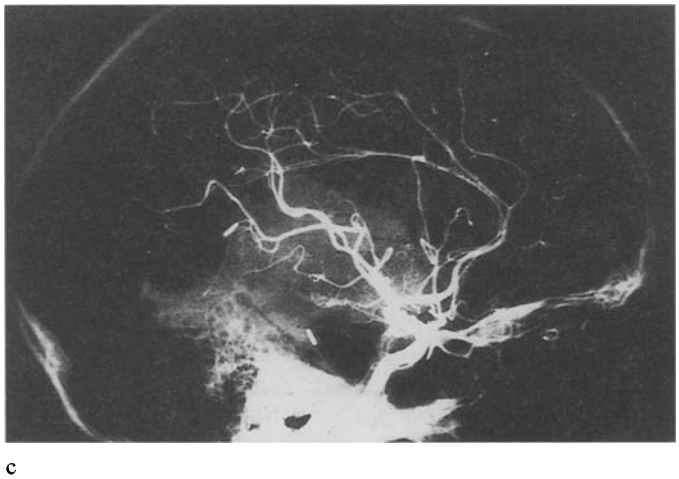

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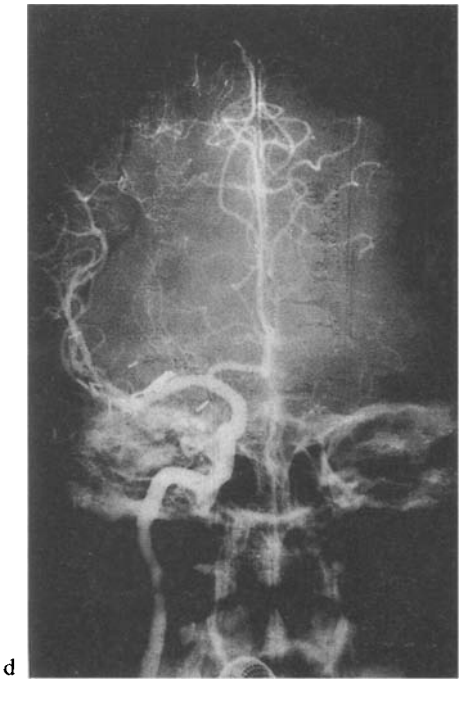
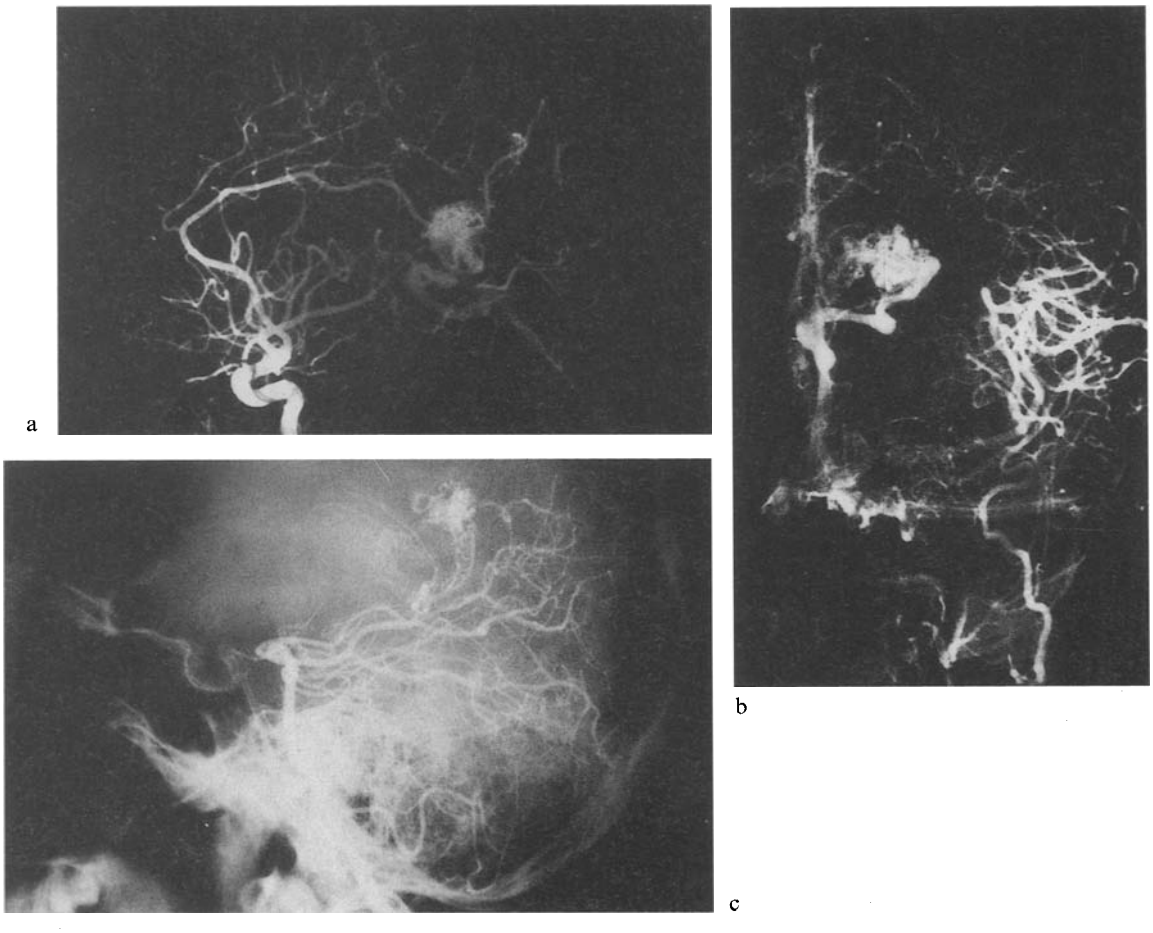


d

Figures 1a—d. Angiograms of AVM in the right Sylvian Fissure. Preoperative figures (**1a** lateral projection, **1b** a. p. projection) show a big venous drainage to the tentorial sinus. Postoperative angiograms (**1c** lateral projection, **1d** a. p. projection) demonstrate successful removal of AVM. (See text, case 1!).



Figures 2a–c. AVM of the splenium of the corpus callosum and the trigone of the left lateral ventricle (**1a** and **b**: preoperative carotid angiograms in lateral and a. p. projection, **2c** preoperative vertebral angiogram). Notice feeders from pericallosal and posterior cerebral arteries. (See text, case 2!).

malformation completely. Seven years later the girl had finished school and is in a good condition with only mild cerebellar ataxia (no figures).

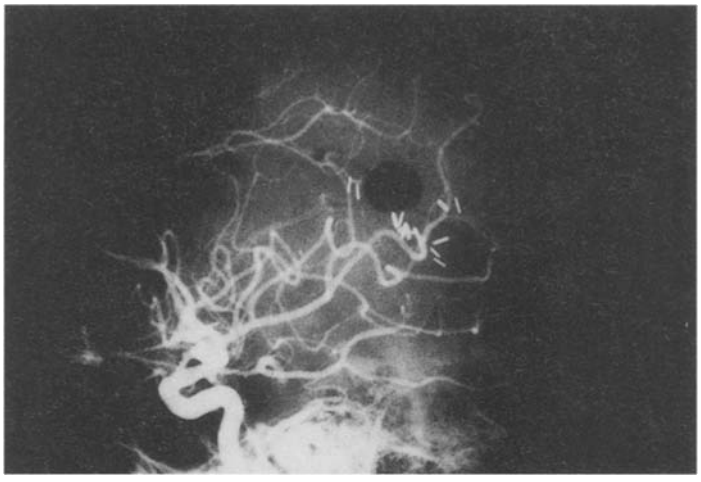
6 Discussion

Arterio-venous malformations are supposed to be the most common cerebro-vascular disorder in childhood [13]. It has been presumed that they are responsible for 30% to 50% of all hemorrhagic strokes in children [1, 6, 11]. However, the exact incidence of AVMs in children as well as in adults is unknown. The prevalence of AVMs in the general population has been calculated to be about 0.14% [14].

545 cases of cerebral arterio-venous malformations and fistulae have been reported in the cooperative study of PERRET and NISHIOKA [15]. 19.6% of the malformations had been diagnosed before the age of 20.

In our personal series of 182 cases treated surgically there were 33 children up to the age of 16 (18.1%) with a slight prevalence of males, similar to the findings of other authors [1, 15].

CELLI and coworkers gave a review of the published clinical data of 242 children up to 15 years reported in the literature. They found the greatest hazard associated with cerebral hemorrhages in children to be a more violent and massive bleeding



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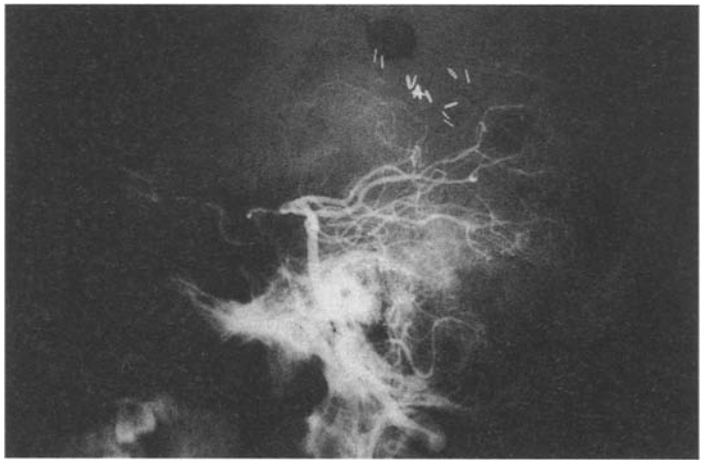

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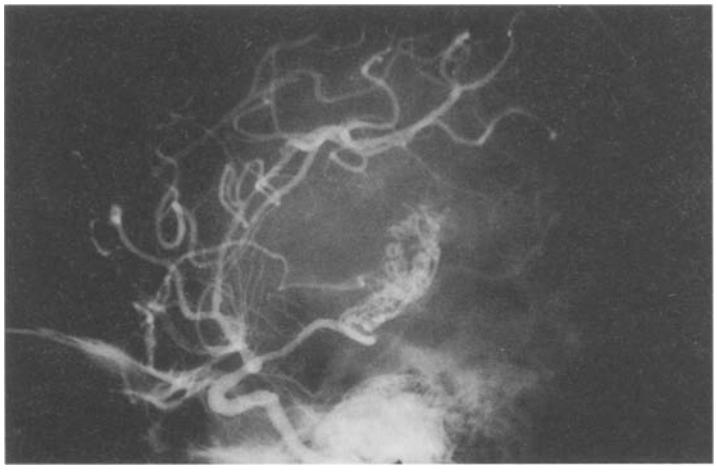
Figures 2d and **e** show postoperative carotid and vertebral angiograms in lateral projection. Total removal of AVM could be achieved.

as demonstrated by the higher frequency of intraparenchymal hematomas and rupture into the ventricles. In our children series, we found a bleeding rate of 84% and intraparenchymal hematomas (clot formation) in 69%. We have already pointed out in a paper published recently [8] that the mode of bleeding in an AVM is influenced by its size and site. We have gained the impression that small and medium-sized angiomas usually bleed early in life whereas large angiomas show a greater variability.

The follow-up studies of patients with AVMs managed without surgery have demonstrated a 42% risk of hemorrhage and a 29% risk of death given a mean of 10.4 years follow-up [2, 4]. On the other hand surgical mortality is about 2.6% in children, compared to 8.5% in adults [1]. These data are comparable to those we got from our own investigations. Because the risk of rebleeding of untreated AVMs seems to increase with advancing age, surgical excision of angiomas should be advised for children even more than for adults. The cumulative bleeding risk of a child with an AVM is higher than that of an adult [1, 5, 7] whereas risks of death from surgery and postoperative morbidity are lower.

As the main purpose of surgical excision of AVMs is prevention of bleeding, there is no controversy as to whether to operate upon AVMs once they have bled. All authors agree that these should be



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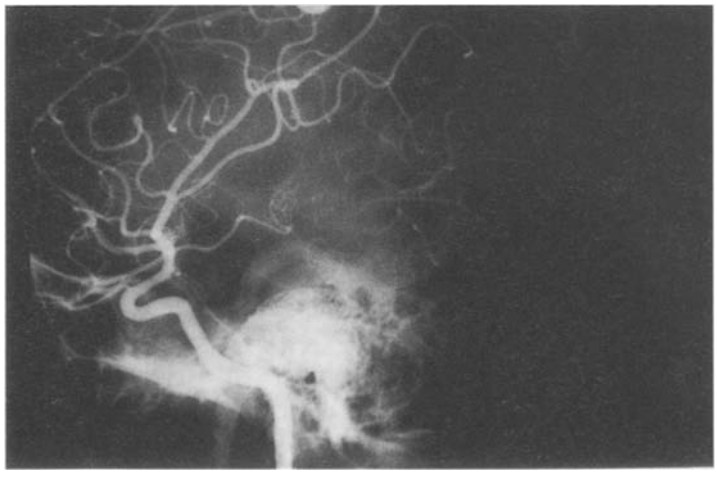

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Figures 3a, b. Preoperative (**3a**) and postoperative (**3b**) carotid angiograms of an AVM of the left choroid plexus. (See text, case 3!).

operated upon [1, 3, 4, 5, 7, 8, 9, 10, 12, 13]. The opinions about angiomas which have not bled are conflicting; especially large angiomas with many arterial feeders and those, which involve a hazardous area of the brain, cause a serious therapeutic dilemma. LUESSENHOP and ROSA developed an anatomical grading system in order to determine the risk of operation of an AVM [12]. They concluded that in the first 3 decades the surgical risk is less than the natural risk for all patients in grades I and II and more than half for those in grade III.

In the current series we observed only one postoperative death in a case with massive intraparenchymal hematoma from a small arteriovenous malformation, whereas the large malformations that had not bled could be removed without any mortality. However, postoperative morbidity was higher than for small and medium-sized angiomas (Table II).

If we do not have any angiographic information about the vascular supply of AVMs in cases operated upon urgently, we prefer to perform a two-stage operation with evacuation of hematoma as a first step and definitive excision of the AVM after recovery of the patient. This procedure has proved to be safe and effective.

7 Conclusions

1. Bleeding is the most frequent symptom of arterio-venous malformations in childhood (85% in our series). Clot formation is much more common than SAH only (with a ratio of nearly 5 to 1 in our series).

2. The cumulative bleeding risk of a child with an AVM will exceed that of an adult, whereas surgical mortality for children is lower than for adults. Surgical excision of AVMs therefore should – once a hemorrhage has occurred – be performed whenever possible.

3. A limited indication towards surgery may be given in large angiomas located in functionally important brain areas without neurological deficit. However, it has to be kept in mind that even after a non-hemorrhagic onset the behaviour of an AVM in further life is unpredictable.

4. Long-term follow-up after removal of AVMs in childhood shows excellent results. Morbidity is comparably low. Most patients of our series were able to reach for their intended occupation.

References

[1] Celli P, L Ferrante, L Palma, G Cavedon: Cerebral arteriovenous malformations in children. Clinical features and outcome of treatment in children and in adults. Surg Neurol 22 (1984) 43–49

[2] Crawford PM, CR West, DW Chadwick, MDM Shaw: Arteriovenous malformations of the brain: natural history in unoperated patients. J Neurol Neurosurg Psych 49 (1986) 1–10

[3] Drake CG: Cerebral arteriovenous malformations: considerations for and experience with surgical treatment in 166 cases. Clin Neurosurg 26 (1979) 145–208

[4] Forster DMC, L Steiner, S Hakanson: Arteriovenous malformations of the brain. A long-term clinical study. J Neurosurg 37 (1972) 562–570

[5] Garza-Mercado R, E Cavazos, D Tamez-Montes: Cerebral arteriovenous malformations in children and adolescents. Surg Neurol 27 (1987) 131–140

[6] Gold AP, YB Challenor, FH Gilles, SP Hilal, A Leviton, EI Rollins, GE Solomon, BM Stein: Report of joint commitee for stroke facilities – IX strokes in children. Part 1. Stroke 4 (1973) 835–894

[7] Graf CJ, GE Perret, JC Torner: Bleeding from cerebral arteriovenous malformations as part of their natural history. J Neurosurg 58 (1983) 331–337

[8] Kahl W, K Dei Anang, G Meinig, M Schwarz: Cerebral angiomas: influence of morphological aspects such as size and site on their clinical behavior with special reference to the mode of bleeding. Neurosurg Rev 10 (1987) 111–115

[9] Kahl W, M Schwarz, K Dei Anang, P Klawki: Intrakranielle Angiome (Erfahrungen an 159 Fällen). Ed Voth D, Neurochirurgia Moguntiaca, De Gruyter, Berlin–New York 1985

[10] Kurokawa T, A Matsuzaki, K Hasuo, M Fukui, S Tomita, M Matsuo, YJ Chen, C Kasemkosolsri: Cerebral arteriovenous malformations in children. Brain Dev 7 (1985) 408–413

[11] Locksley HB: Report on the cooperative study of intracranial aneurysms and subarachnoid hemorrhage. Section V, Part 1, Natural history of subarachnoid hemorrhage, intracranial aneurysms and arteriovenous malformations. J Neurosurg 25 (1966) 219–239

[12] Luessenhop AJ, L Rosa: Cerebral arteriovenous malformations. Indications for and results of surgery and the role of intravascular techniques. J Neurosurg 60 (1984) 14–22

[13] Matson DD: Neurosurgery of infancy and childhood. 2nd ed. Charles C Thomas, Springfield, Ill. 1969 749–766

[14] Michelsen WJ: Natural history and pathophysiology of arteriovenous malformations. Clin Neurosurg 26 (1979) 307–313

[15] Perret G, H Nishioka: Report on the cooperative study of intracranial aneurysms and subarachnoid hemorrhage. Section VI. Arteriovenous malformations. An analysis of 545 cases of craniovertebral arteriovenous malformations and fistulae reported to the cooperative study. J Neurosurg 25 (1966) 467–490

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