ANATOMICAL AND CLINICAL ASPECTS OF CEREBRAL ARTERIOVENOUS MALFORMATIONS

N.C. Florestean[1], A.V. Ciurea[2]

[1]Emergency Department, "Bagdasar Arseni" Clinical Emergency Hospital, "Carol Davila" University of Medicine and Pharmacy, Bucharest

[2]Neurosurgical Clinic, "Bagdasar Arseni" Clinical Emergency Hospital, "Carol Davila" University of Medicine and Pharmacy, Bucharest

ABSTRACT

This article describes the anatomical and clinical aspects of cerebral arteriovenous malformations which from the start won the role of an important cause of death or severe morbidity in the long term due to intracranial hemorrhage and secondary epilepsy. It is the second leading cause of aneurysms after subarachnoid hemorrhage. Cerebral arteriovenous malformations present a nidus fed by one or more nourishing arteries ("feeders") with deep or superficial venous drainage. Symptoms are determined by the presence of seizures and intraparenchimatous and/or subarachnoid bleeding. Progressive neurological deterioration can exist, which is attributed to ischemic disorders due to "vascular theft ". Also, it is described the grading system of arteriovenous malformations depending on size, eloquence and venous drainage by Spetzler-Martin system. Also described, a number of other scales with prognostic role and evaluation of the patient's consciousness, but in was in 1988 when the World Federation of Neurological Surgeons proposed a simplified scale, based on clinical findings of Hunt and Hess scale, translated into GCS, WFNS grading scale SAH.

Key words: cerebral arteriovenous malformations (C-AVM), nidus, subarachnoid hemorrhage (SAH), Hunt and Hess scale, Scale GCS, scale GOS, the Spetzler-Martin system, WFNS scale

Since their first description more than a century ago (Steinheil, 1895) (7) cerebral arteriovenous malformations (C-AVM) have gradually gained the role as a significant cause of death or severe long-term morbidity, primarily due to intracranial hemorrhage and secondary epilepsy. The technological development of vascular brain imaging systems and increase of their addressability (CT, MRI and DSA) increased the rate of diagnosis of C-AVM (2), so that now the diagnosis can be made as a routine activity. There is a growing interest regarding the incidence and clinical course of C-AVM, especially since the development of microsurgical, endovascular or radiosurgical treatment methods have improved the prognosis of the patients with C-AVM. The individual prognosis still retains a high degree of uncertainty, as well as the risks and benefits of various treatment methods, which leads to large variations in therapeutic attitude and hence to the need for randomized clinical trials.

Developing some accurate criteria for the diagnosis of C-AVM and defining as accurate as possible the vascular anatomy variants, the so called angio-architecture, are essential in determining the criteria for developing the clinical trials and then in the correct use of the collected data in the future patients' treatment planning. The role of brain imaging in the diagnosis of C-AVM is especially important because the study of angio-architecture leads to establishing the patient's prognosis, both in respect of the natural evolution of the disease and of the results of various applicable treatment methods.

Cerebral arteriovenous malformations (C-AVM) represent, after aneurysms, the second cause of subarachnoid hemorrhage (SAH), in children they

Author for correspondence:
N.C. Florestean, "Carol Davila" University of Medicine and Pharmacy, 37 Dionisie Lupu St., Bucharest

even coming on the first place. Their presence is manifested most often by crisis, but also by often massive intra-parenchymal hemorrhages, which can sometimes lead to death (Fig. 1).

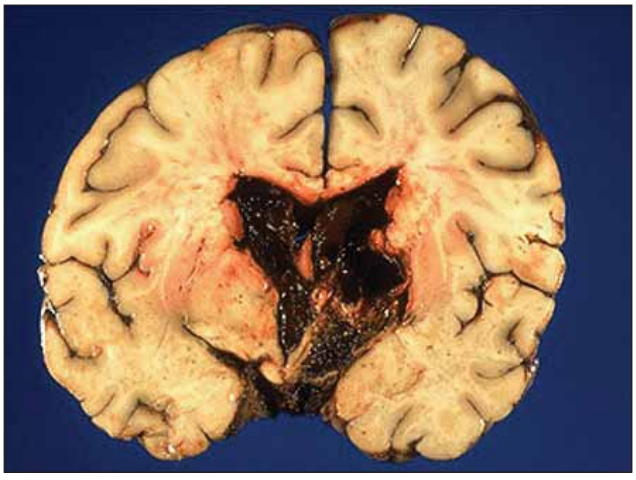

FIGURE 1. *Broken C-AVM with cerebral hemorrhage in basal nuclei and intraventricular effusion (4)*

Another physiopathological consequence of the AVMs is the "steal" syndrome, clinically characterized by motor deficits or sometimes by heart failure secondary to a venous return in excess, more common in children. C-AVM are part of hamartomas group, they establishing a massive arteriovenous short-circuit, with deprivation of capillary circulation from the relevant territory and from the surrounding territory of normal blood supply. They are formed around the third week of intrauterine life, when the large vessels appear, the arteriovenous communication substituting itself to the capillary bed development.(1)

C-AVMs usually occur with a "nidus" fed by a variable number of arteries ("feeders") and drained by a series of hypertrophied veins (Fig. 2).

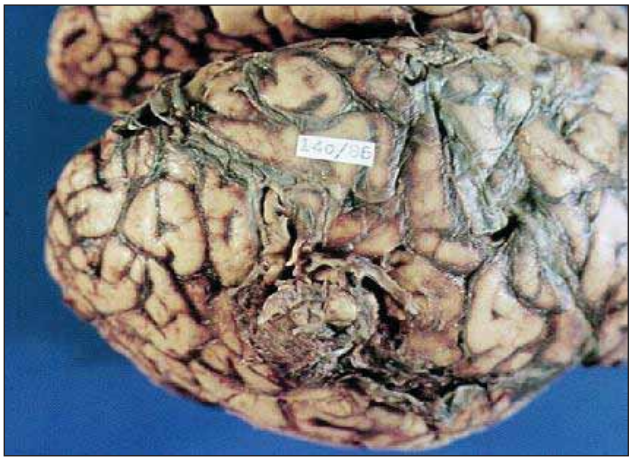

FIGURE 2. *Corticalized Nidus P-O (anatomic preparation) (4)*

Inside the nidus there are usually found cavernous structures that can not be classified either as veins or as arteries; in the surrounding parenchyma hemorrhagic foci can be observed with macrophages and disappearance of neurons, but also ischemic areas. The vessels have many parietal hyalinisation lesions and thrombosis. The often para-ventricular location of the nidus explains the frequency of intraventricular effusions, in the case of these breakings. The most frequent arterial sources are located at the level of the middle cerebral arteries, followed by anterior cerebral arteries and then by the posterior cerebral arteries. Often the nutritious arterial branches pass through a tortuous paths at the level of the sulcuses, before entering the nidus. The nervous tissue surrounding the nidus is gliotic and dysfunctional. Sometimes, however C-AVM appears diffuse, with normal parenchyma within it, making it harder to be removed. The overall incidence of C-AVMs is still uncertain, Mc Cormick estimating it at 0.5% (4). Generally, it is accepted that the frequency of C-AVM represents 50% of the frequency of aneurysms.

CLINICAL ASPECTS

The symptoms, as mentioned above, are determined by the presence of crises and bleeding. Crises usually take a focal aspect. Hemorrhages are intraparenchimatous, they externalizing, however, also in the subarachnoid spaces. In general, the bleeding, as well as bleeding recurrence rate are less devastating than in the case of the aneurysms. Their rupture is, however, most common in children. The progressive neurological deterioration can also be found without hemorrhage, they being attributed to ischemic disorders due to "steal". Besides headache often of migraine type transient amblyopies, papillary edema, assigned to intracranial venous hypertension, can be cited. These "pseudotumor cerebri" phenomena usually disappear after removal of malformation.

The natural history of C-AVMs is difficult to ascertain due to high variability of their symptoms. In larger or lesser series a mortality of about 25% within 12-24 years was recorded. It was found in the same series a bleeding recurrence rate of about 4% per year and a morbidity combined with a mortality of about 2.7% per year (Fig. 3). However, about 68% of these patients managed to lead a relatively normal life. Mortality after the first bleeding of a C-AVM is about 10% and for the next bleedings it reaches 20-40%. In children under 15 years, the prognosis is better. As elements that would fa-

vor the rupture the following could be mentioned: small size, location and high hemodynamic resistance. The emergence of embolization techniques combined with selective approach of some superficial lesions of limited size, occurring in young people gave a new impulse for these lesions to be attacked in a pre-hemorrhage phase. (5)

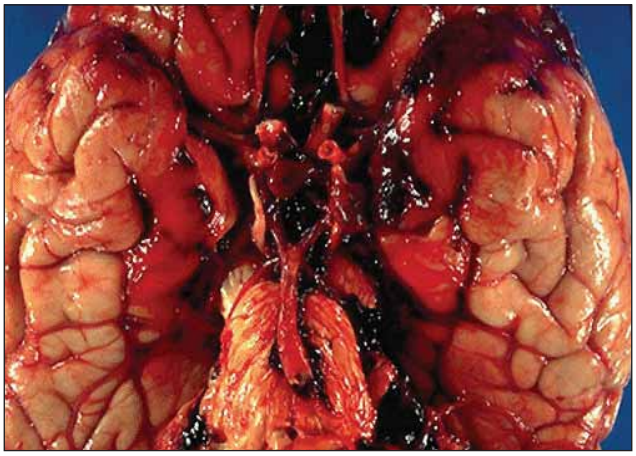

FIGURE 3. *Subarachnoid hemorrhage secondary to the rupture of a BAVM of basal nuclei.*
(Report of World Federation of Neurological Surgeons Committee on an Universal Subarachnoid Hemorrhage Grading Scale)(5)

There were also quoted some sporadic cases of spontaneous regression of even larger lesions, but having a single supply source and a drainage source. Such spontaneous regressions appear to be related to the occurrence of some bleeding episodes, to irradiating treatments or to a thrombotic spurt, often revealed by neurological deficits.

Tracing the lesions by means of repeated angiographies for periods between 5 and 28 years showed the following trends: 20% increase in size, 20% extinctions and 40% remained stationary. Recurrent bleedings of 35% were noted in this series. In general, the increase in size was recorded in young people and reduction in older people. Spontaneous regression occurs only exceptionally and it belongs of course to the tendency to progressive fibrosis and vascular hypercoagulability of the person in question. In general, however, C-AVM evolution translates into ischemic outbreaks occurring after haemorrhage and causing neurological deficits. (2)

C-AVM GRADING SYSTEMS

C-AVM grading systems were established to create standardizing parameters for irrigation, size, location and drainage of malformation, in order to establish a rate of morbidity and mortality for each case. This system, proposed by Spetzler and Martin (Table I), is the most used; it has six degrees, each referring to the size, location and venous drainage (6). The size is measured at the maximum angiographic diameter of C-AVM.

The location, taking into account the function of brain area where C-AVM is located, will mark the seat in the senzorimotor area, in the language or visual cortex area, in the diencephalon, internal capsule, brain trunk or deep cerebellar trunk.

The venous drainage raises special problems when the galenic system is involved in the drainage. In the cases fitting in grade III or in a higher grade, high operative morbidity is likely to occur. In the case of grade V, operative mortality can also occur, and the cases fitting in grade VI are considered inoperable. Preoperative embolization as well as the partially seriated operation can reduce the surgery risk grade.(1)

Thus, this clinically relevant classification, integrating the knowledge about C-AVM morphology with etiological data known at this time and the differences in prognosis and response to various treatment methods is very helpful in differentiating C-AVM from other intracranial venous malformations. This difference can be made by appropriate use of the methods of invasive and noninvasive imaging of cerebral vascular tree.

Due to the extreme variety of presentation, location and angio-architecture of C-AVM, Spetzler and Martin proposed in 1986 (6), based on the analysis of their personal experience with 100 consecutive cases, a prognostic scale that has proven its validity over the years and that also became, through international recognition, an extremely practical method of C-AVMs classification.

TABLE 1. *Spetzler-Martin grading system of C-AVM (6)*

Feature	Points
Size (maximum diameter):	
small (< 3 cm)	1
medium (3-6 cm)	2
large (> 6 cm)	3
Eloquence of adjacent brain parenchyma (sensorimotor, language, visual etc. cortex, hypothalamus and thalamus, internal capsule, brainstem, cerebellar peduncles, deep cerebellar nuclei):	
non-eloquent	0
eloquent	1
Venous Drainage:	
superficial (cortical veins only)	0
deep (any of internal cerebral veins, basal veins or pre-central cerebellar veins)	1

Spetzler-Martin grade of C-AVM is given by the sum of points awarded based on the characteristics listed in Table 1. Thus, a small C-AVM located in non-eloquent area of the brain and with superficial venous drainage only will Spetzler-Martin Grade 1, while a C-AVM larger than 6 cm in diameter located in an eloquent area and with deep venous drainage will be Grade 5. A special category that received "ex officio" the Grade 6, is the C-AVM unapproachable by any of the currently available therapeutic methods (microsurgery, radiosurgery or endovascular surgical approaches), the risk of death or severe neurological deficits being unacceptable high.

TABLE 2. *Hunt and Hess scale*

Grade 0	C-AVM/untorn aneurysm
Grade 1	Asymptomatic patient or mild headache or moderate neck stiff
Grade 1a	Patients without meningeal or brain reaction but with acute neurological deficit
Grade 2	The patient shows paresis of cranial nerves (III, VI), moderate or severe headache, neck stiff
Grade 3	The patient is drowsy, confused and shows minimal neurological deficits
Grade 4	The patient is comatose with moderate or severe hemiparesis with decerebration rigidity and vegetative disorders
Grade 5	The patient is in deep coma, with decerebration rigidity, dying

Adapted from Hunt W., Hess, R., J. Neurosurg 1968; 28:14 (3).

TABLE 3. *Glasgow Coma Scale*

	Points
Eye Opening	
Spontaneous	4
In response to verbal commands	3
In response to painful stimuli	2
Does not open the eyes	1
Verbal response	
Oriented	5
Confused	4
Inappropriate words	3
Incomprehensible sounds	2
No verbal answer	1
Motor response	
Responds to commands	6
Locates pain	5
Withdraws the limb to painful stimuli	4
Reflex response in flexion to painful stimuli	3
Reflex response in extension to painful stimuli	2
No motor response	1

Teasdale G., Jennette B. Assessment of coma and impaired consciousness. A practical scale. Lancet 1974; 2:81 (8).

TABLE 4. *Glasgow Outcome Scale*

Patient's condition	Category
Good recovery	5
Moderate disability (with sequels, but independent)	4
Severe disability (conscious, but with serious sequelae)	3
Persistent vegetative condition	2
Died	1

Subarachnoid hemorrhage cerebral (SAH), or intra-parenchymal hemorrhage cerebral (IPH), is the strongest form of onset or complication in the development of cerebral arteriovenous malformations. Hence, the constant concern of neurologists doctors and neurosurgeons to develop prognostic scales as reliable, simple and generally applicable as possible, for their use in therapeutic decision making optimal algorithm before a patient with cerebral hemorrhage with an aneurysm or vascular malformation origin. Hunt and Hess scale (3) (shown in Table 2) was one of the first coherent, enough simple and reproducible scales, with proven prognostic value over time. For these reasons, also now, almost 40 years after its invention, it is still the most commonly used clinical grading scale for patients with SAH.

With the widespread use of Glasgow Coma Scale "GCS" and Glasgow Outcome Scale "GOS" (6) (other extremely simple, reliable and easily reproducible scales including by medical personnel with basal training, presented in Tables 3 and 4) in all patients with impaired consciousness (not only in the head trauma), it became necessary the various clinical grading systems for patients with SAH, to be "unified", based on GCS. Thus, it was proposed by the "World Federation of Neurological Surgeons" (Table 5) in 1988 (5) a simplified scale, based on clinical findings of Hunt and Hess scale, translated into GCS.

TABLE 5. *SAH grading WFNS scale*

WFNS grade	GCS Score	Motor deficit
I	15	Absent
II	14-13	Absent
III	14-13	Present
IV	12-7	Present or absent
V	6-3	Present or absent

Adapted from: Report of World Federation of Neurological Surgeons Committee on an Universal Subarachnoid Hemorrhage Grading Scale. J Neurosurg 1988; 68:985 (5).

REFERENCES

1. **Brown R.D., Jr., et al.** – Incidence and prevalence of intracranial vascular malformations in Olmsted County, Minnesota, 1965 to 1992. *Neurology,* 1996. 46(4): p. 949-52.
2. **Constantinovici A., Ciurea, A.V.** – Tratamentul malformatiilor arterio-venoase ale SNC. Ghid practic de neurochirurgie. 1998 p. 379-380
3. **Hunt W.E. and R.M. Hess** – Surgical risk as related to time of intervention in the repair of intracranial aneurysms. *J Neurosurg,* 1968. 28(1): p. 14-20.
4. **McCormick W.F.** – The pathology of vascular ("arteriovenous") malformations. *J Neurosurg,* 1966. 24(4): p. 807-16.
5. Report of World Federation of Neurological Surgeons Committee on a Universal Subarachnoid Hemorrhage Grading Scale. *J Neurosurg,* 1988. 68(6): p. 985-6.
6. **Spetzler R.F. and N.A. Martin** – A proposed grading system for arteriovenous malformations. *J Neurosurg,* 1986. 65(4): p. 476-83.
7. **Steinheil S.O.** – Ueber einen Fall von Varix aneurysmaticus im Bereich der Gehirngefaesse. Wiirzburg: F. Fromme, 1895: p. 1-56.
8. **Teasdale G. and B. Jennett** – Assessment of coma and impaired consciousness. A practical scale. *Lancet,* 1974. 2(7872): p. 81-4.