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Personal Experience in the Treatment of 178 Cases of Arteriovenous Malformations of the Brain

By

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With 4 Figures

Summary

We have analysed our personal series of 178 consecutive cases of arteriovenous malformations (AVMs) treated since 1954. A surgical approach was used in 146 cases (820/0), with direct attack on the malformations in 132 cases (740/0). There was total extirpation of the malformations in 124 of the operation cases (940/0). In the remaining cases, ligation of the carotid artery was performed in three, and ligation of the afferent feeding artery in five. Embolization was used in five cases, and stereotaxic cryocoagulation in one.

The decision-making process was based on the size of the malformations and the clinical presentation, which was subarachnoid haemorrhage (SAH) in $64^{0}/o$ of the cases, and epilepsy in $15^{0}/o$. The results were considered good in 117 of the cases ($89^{0}/o$), and the mortality rate was $11^{0}/o$. As in other series, a high incidence of AVMs was encountered in patients under 40, and men predominated ($74^{0}/o$) over women.

Different criteria of operability are discussed, including a "New Look" at the venous components of the malformations as an important assessment.

Keywords: Arteriovenous malformations; surgical procedure; criteria for operability.

We treated and observed 178 cases of AVMs of the brain from 1954 until November 1980. The progressive increase in number of cases is very evident in our material, which comprised 40 cases from the period of 1954 to 1966, 60 cases from 1966 to 1974, and 78 cases from the period of 1974 to November 1980. Because of the number of cases, we no longer consider these lesions as infrequent, and we treat them as a benign intracranial processes, with the potential to bleed, and with a similar incidence as, for instance, acoustic neurinomas.

Definition

When speaking of this subject we have repeatedly insisted on the necessity to abandon the old classifications of cerebral vascular malformations which, from our point of view, only create conceptual and therapeutic confusion. The unitarian diagnosis of all these malformations must be based on the arteriographic objectivity of the arteriovenous communication, which almost always is of a congenital nature, and may be enormous. Large efferent venous dilatations can conceal the real arteriovenous fistulas. Only a serial arteriographic study, in rapid sequence, can distinguish the true fistula, whether it be large, small, simple, or complex, that should be the only object of our surgical attention. Its occlusion or destruction restores the normal circulatory physiology of the brain, which is the goal of all therapeutic procedures.

Own Material

Location and First Symptoms

As in other published series $^{3.17-19.24-28}$ the supratentorial AVMs have much greater frequency than those located in the posterior fossa (in ours 168 and 10 cases respectively, Table 1), with a high number of AVMs of the midline of paramedial areas, that is to say, those affecting the corpus callosum, the basal ganglia, and the brain stem (35 cases). The same can be said in respect of malformations, when the external carotid is involved. They form a group of 14 of our cases, with well-defined clinical characteristics and therapeutic problems different from those of the internal carotid system. Table 2 embodies the presenting symptomatology of all our cases. Subarachnoid haemorrhage is without a doubt the most frequent presenting feature, followed by convulsive crises and the steal phenomenom.

Age and Sex Incidence

When observing the age and sex of the patients, it was noted that there was a higher incidence of AVMs in the first decades of life in both sexes, but there were twice as many males as females.

Diagnostic Procedures

Only angiography gives a precise diagnosis of the location, extent, and vascular feeding and draining of a malformation. The value of these data, along with the clinical findings, is to allow the employment of the best therapy for each case. The angiographic study must embrace both the carotid systems and the vertebro-basilar system. Even when the clinical symptomatology allows the suspicion that the site of the lesion is in a cerebral hemisphere, we must not limit ourselves to doing the angiography on one side only, since a complete study sometimes discloses multiple malformations and, above all, it shows us that a malformation may receive arterial feeding from several homo- and contralateral systems. The AVMs situated in the anterior halves of the cerebral hemispheres are usually fed by the

	frontal	29
	parietal	44
Supratentorial (hemispheres)	temporal	25
	occipital	13
	Sylvian	8
	corpus callosum	11
Midline and paramedial	basal ganglia	22
^	brain stem	2
* C	vermis	3
Infratentorial	cerebellum hemispheres	7
AVMs with the participation	of the external carotid	14

Table 1. Location Topography of the A	VMs
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	Number of cases	
Subarachnoid haemorrhage (with or without haematoma)	115	65%
Convulsive crisis	26	15º/e
Steal phenomenon	23	13º/o
Endocranial hypertension syndrome	4	2º/o
Headaches	6	3%/0
Psychic deterioration	4	2º/o

Table 2. Presenting Features

Sylvian group, the homolateral anterior cerebral system, and frequently by the contralateral system, and when they are deep-seated they also receive feeding from the thalamo-striate artery and the anterior choroidal artery. The AVMs situated in the posterior half of the cerebral hemispheres and the basal ganglia, besides being fed by the middle cerebral and anterior cerebral systems, are fed by the posterior cerebral system, which almost always depends on the vertebro-basilar system and posterior choroidal arteries (Fig. 1).

This multiplicity of arterial feeding to the malformation is due to its embryonic origin. In general, this has led us to consider the

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ineffectiveness of surgical methods of ligation of the afferent vascularization, and inclines us towards the direct extirpation of the lesion as being the only efficient treatment. If, in control angiographs, a complete study is not made, after removing an angioma errors can be made, by supposing that it has been completely extirpated, when in fact parts of it are merely not visible and are depending on feeding vessels other than those shown by simple homolateral angiography.

In our cases the AVMs situated in the anterior part of the hemispheres were fed basically by the Sylvian arteries in $55^{\circ}/_{0}$, while in $16^{\circ}/_{0}$ blood also came from the ACA or one of its branches. In $21^{\circ}/_{0}$ of our cases of AVMs situated in the occipital and basal ganglia both the carotid and vertebro-basilar systems were responsible for AVM circulation. In the 10 cases of posterior fossa AVMs, the right vertebral artery was involved in 6 cases and the left vertebral artery in the other 4. Verbiest stresses the possibility of carotid artery participation in posterior fossa AVMs³⁰, but like him we have not found any cases of this.

In the literature there is much controversy about the importance of the drainage system, not only in respect of technical management, as Malis¹⁷ has pointed out, but because the direction of the contrast flow is reversed in patients with a low bleeding risk, as Pellettieri *et al.*²⁴ found. The idea of blood distribution in many directions, diminishing the intravascular "traffic-like" pressure is tentative.

We have studied 30 patients, trying to assess whether there was a clear relationship between the number of drainage veins and the initial symptoms, and found that, when the malformation has one or two venous drainages, the first feature is haemorrhage, and in cases where there were three or more venous drainages, the principal initial feature was epilepsy or mental retardation. We feel that these are important facts, which could have prognostic value when deciding whether or not to operate. We are now considering reviewing our case material with this point of view.

The small (2 cm or less) arteriovenous malformations usually have few drainage veins, even if these appear large, and they have the tendency to bleed more ³⁴ than the medium or large ones. Jos *et al.* ¹² came to the same conclusions; they reviewed the so-called microangiomas in our material and found that in 90% the initial feature was subarachnoid haemorrhage.

In recent years CAT, as in other types of intracranial pathology, has contributed some very interesting information. Sometimes, without images that can be considered pathognomonic of a vascular malformation, CAT can indicate an AVM by its morphology and venous drainage, and together with clinical data can suggest the aetiology of



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the lesion in some cases of epilepsy and subarachnoid haemorrhage. In bleeding malformations, it can give the precise location and extent of intraparenchymatous or intraventricular haematomas, and show whether ventricular dilatation exists or not. At other times, it shows the atrophied cerebral area encircling the malformation, which is a consequence of repeated haemorrhages or of anoxia of the surrounding cerebral area due to the steal phenomenon.

Treatment

We have already mentioned that we consider the total removal of a malformation as the only efficient treatment. We have done this in a very high percentage of cases. The surgical procedures employed

		Results				
		Good	Fair	Bad	Deaths	
D' 1	radical extirpation	105	2	2	15	
Direct attack	partial extirpation	4	3	1		
	cervical carotid	3				
Arterial ligations	afferent artery	3	2			
Embolization		2	2		1	
Cryocoagulation				1		
	Totals	117	9	4	16	

Table 3. Surgical Techniques and Results (patients operated on: 146)

in the treatment of our patients are shown in Table 3. Of the 178 patients we observed, 146 were operated on. Of these 146 patients, 124 had radical extirpation $(85^{0}/_{0})$, and a partial removal was done on 8. The afferent arteries of eight patients were treated by means of ligation, five were embolized, and only one patient was treated by cryocoagulation.

Some of the largest series published in recent years are shown in Table 4. They indicate the relation between the number of cases studied, the number of patients operated on, and the mortality rate.

In Table 5 we show some of the series found in the bibliography that specify the number of cases in locations that could be called problematical and that were considered, until a short time ago, as being unapproachable by most authors. In our studies, these locations stand out as being more frequent than others, and yet the percentage of surgical treatment is also greater. Without the mortality associated

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with these locations, the overall mortality rate, in almost all of the series, would be lower. We think that in the presence of AVMs, particularly those in deep-seated locations, neurosurgeons suffer from a general inhibition of aggressiveness which is not rational and is based on the experience based on the techniques of their time transmitted by the pioneers of neurosurgery, such as Olivecrona²², Tönnis²⁹, and Krayenbühl⁶, who considered these areas as being in-

No. of cases	No. of cases operated on	Surgical mortality for radical extirpation
150	44 (30%)	70/0
125	81 (65%)	9%/0
?	123	14º/o
118	68 (58 ⁰ /0)	9º/o
154	102 (66%)	8°/0
205	165 (80%)	14%/0
196 (174 supratentorials)	98 (56 ⁰ /0)	4º/0
166	119 (72%)	13%/0
?	162	11%/0
?	166	13%
178	146 (82%))	11%
	No. of cases 150 125 ? 118 154 205 196 (174 supratentorials) 166 ? ? 178	No. of cases operated on 150 44 (30%)(0) 125 81 (65%)(0) ? 123 118 68 (58%)(0) 154 102 (66%)(0) 205 165 (80%)(0) 196 98 (56%)(0) (174 supratentorials) 166 166 119 (72%)(0) ? 162 ? 166 178 146 (82%)(0)

Table 4

operable. We believe that, if this inhibition ceases all well-trained neurosurgeons knowledgeable in microsurgical techniques, and working in an adequate neurosurgical environment, can and must move towards a low mortality rate.

Microsurgery is a positive advance in the treatment of cerebral AVMs. Before microsurgery, we were followers of extirpation by means of destruction by coagulation and aspiration of the nucleus of the angioma, almost without the use of haemostatic clips. In our experience with macrosurgical techniques, there is much less loss of blood, and the operation is more rapid than when one performs vessel by vessel occlusion of the angioma, predominantly with clips. Actually, microsurgery, using bipolar coagulation, permits a regulated and bloodless extirpation of a large part of an AVMs, and it allows a much longer operating time, that does not affect the excellent final results. However, experience has taught us that microsurgical tech-

Table 5. Authors

Morello and Borghi

Cases studied

Cases operated on 102 Cases Operated on Basal ganglia 12 1 Mesencephalon 8 1 Ponto-cerebellar 3 1 Corpus callosum 0 0 Cerebellum 0 4 $7 (22^{0}/_{0})$ 31 Mortality unknown Sano et al. Cases studied 205 Cases operated on 165 Cases Operated on Paraventricular Basal ganglia 30 $6 (20^{\circ}/_{\circ})$ Brain stem Mortality unknown Pertuiset et al. Cases studied unknown Cases operated on 162 Operated on Cases Basal ganglia 6 Intraventricular septum 1 Galenic cistern 1 Cerebellopontine angle 6

Mortality unknown

154

166

Table 5 (continued)

Drake, C. H.

Cases studied	
Cases operated	on

	Cases	Operat	ed on	
Corpus callosum Intraventricular		6 (1 5	death)	<u> </u>
Basal ganglia		5		
Cerebellum		14 (1	death)	
Brain stem Cerebellopontine		4 (2	deaths)	
				<u></u>
		40		
Mortality		5 (1	.3º/0)	
Albert <i>et al.</i>				
Cases studied				178
Cases operated on		146		
	Cases	Operat	ted on	
Basal ganglia	22	15 (2	2 deaths)	
Brain stem	2	0 Ì	,	
Corpus callosum	11	10 (1	l death)	
Cerebellum	10	10		
	45	35 (2	78%/0)	
Mortality	<u> </u>	3 (9	90/0)	
In these cases radical extirpation	ns were made in	26 (58º/o)	

niques are difficult to apply to malformations or areas of malformations that present the classical spongy or plexiform structures, and for these we recommend employing the new techniques until the control of haemorrhage is no longer possible, and then one should continue with the old technique of aspiration and destruction of the

unknown

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malformation. Microsurgery has expanded the field of indications considerably by allowing us to approach deep-seated malformations through routes of limited access and with perfect illumination. The angiomas of the corpus callosum, fornix, foramen of Monro, splenius, region of the Galen veins, basal ganglia, and the cerebellum are the principal areas for the new technique.

The transcallosal pathway (Fig. 2) to many of these malformations has shown itself to be very useful and without apparent sequelae from the callosotomy. This observation was made recently by

			Good	Fair	Bad	Deaths
Corpus callosum	not operated on operated on (direct attack	1				
(II cases)	and radical extirpation)	10	9			1
	(not operated on	7				
Basal ganglia	operated on: embolization	1		1		
	ligation of afferents	2	1	1		
(22 cases)	stereotactic	2	1	1		
	direct attack	10	7	1		2
Brain stem (2 cases)	not operated on	2				

Table 6. Midline and Paramedial Arteriovenous Malformations (Surgical Techniques and Results)

Yaşargil^{35, 36} and Drake³, and in 15 patients in our Department normal activity followed. In one case we performed a total callosotomy to remove an AVM that extended almost to the corpus callosum. The patient, a male nurse, operated on ten years ago, is now working normally. The neuropsychological tests and visual evoked potentials are of great importance to this subject, and should be used in preand postoperative studies. They are of great value in investigating the safety of this approach. The callosotomy and the opening of the corresponding lateral ventricle allows the visualization in the anterior part of the fornix, foramen of Monro, the head of the caudate nucleus, and in the posterior part of the thalamus, the choroid plexuses, the splenium, the pulvinar, and the Galen vein area, besides allowing access to the contralateral ventricle, if necessary. This midline route, not using cortico-subcortical tunnels, does produces neither epilepsy nor hemianopias, as when entry is made through the temporo-parieto-occipital cortex (Table 6). These AVMs are fed



Fig. 2. Total removal of a splenium AVM throught a transcallosal approach

by the choroidal arteries, the perforating branches, and thalamo striated arteries, and there is generally less bleeding involved when extirpating them than in the rest of the hemispheric angiomas in which the Sylvian artery fundamentally participates.

Our experience has been satisfactory: in eleven cases of AVMs of the corpus callosum, radical extirpation was performed in nine and they have survived without sequelae. One patient refused operation, and the only death was in a woman in her fifth month of pregnancy, on whom an emergency operation was performed for intraventricular haemorrhage. She had a subcallosal angioma in the roof of the third ventricle.

As to the AVMs of the basal ganglia (Fig. 3), the possibilities of removal are quite diverse; the small paraventricular ones which generally bleed a lot and whose extirpation is easy, are difficult because of their locations. In some of these, stereotactic procedures have been employed in order to localize them correctly. The angiomas of the posterior thalamus almost always extend towards the region of the splenium, and are completely approachable through the transcallosal route. On the other hand, certain large angiomas of the thalamus continue to be unapproachable. Of our 22 cases, intervention was not indicated in 7, and of the remaining 15, radical extirpation was performed on 5 that had been bleeding and which had intraparenchymatous haematomas. There were two deaths. In the other two large angiomas, we limited ourselves to ligating the afferent arteries, and the result in one patient was excellent and in the other fair. We have only performed intraoperative selective embolization on one internal capsular angioma, using intraoperatory anteriographic control. The clinical results were good, but there was a persistent remnant of the malformation in the final angiographic examination.

The angiomas of the posterior fossa. In this group, most authors include angiomas of the cerebellum, the brain stem, and those which affect both structures, and consider them to be hardly likely to respond to surgical treatment. Verbiest ^{30, 31} also includes in the same group dural angiomas, and what he calls subarachnoid angiomas which are located in the cistern of the cerebellopontine angle or in the region of the Galen veins.

Our practice is to make a clear separation between angiomas with predominant participation of the external carotid artery draining to the dural sinuses and the rest of the AVMs. In this group (Table 7) we deal exclusively with angiomas of the cerebellum, whatever their size or location (Fig. 4).

AVMs dependent on the external carotid artery, in our experience, constitute the group with the most treatment difficulties. Within this



Fig. 3. Left basal ganglia AVM with intraventricular haematoma. Total extirpation

Table 7. Posterior Fossa AVMs

Infratentorial AVMs	10
Operated on	10
Total extirpation	10
Vermis	3
Hemispheric	7
Mortality	0

group, the cases in which the external carotid artery contributes only a small amount to the vascularization of the angioma and which basically depend on the internal carotid should be put to one side. They constitute only a small number of our case material. Funda220

mentally, we refer to a certain type of malformation in which several branches of the external carotid artery and the extracranial branches of the vertebral flow into the dural sinuses, generally into the posterior part of the longitudinal sinus and into both the transverse and sigmoid sinuses, being able to produce large arteriovenous lakes between the two layers of the tentorium (Verbiest group III 30). They also receive vascularization from the anterior and posterior meningeal arteries. Clinically they produce very severe symptomatology, owing to the massive steal phenomenon, with mental deterioration, convulsive crises, transitory paresis, and intracranial hypertension. The increase of pressure in the cerebral venous drainage system explains the intracranial hypertension and, in general, the gravity of these fistulas. A direct attack on these malformations is not possible because of their large size, and their treatment is limited to ligating or embolizing the feeding arteries to the dural sinuses. It is feasible to do this bilaterally, using the branches of the external carotid artery above the lingual artery. It is more difficult to obliterate the muscular branches of the vertebral artery, and the exclusion of the meningeal arteries is practically impossible, especially exclusion of the meningeal artery of the tentorium (the Bernasconi artery), which also participates in these malformations. Many arterial afferents, not visible in the first angiographic studies, hypertrophied, and manifested themselves when the arteries dependent on the external carotid artery were excluded, and this often made our efforts useless.

We have already mentioned that, in general, *ligation of the* afferent arteries is ineffective, and that it is only indicated when it has been demonstrated with certainty by arteriographic studies that the pedicles to be occluded are the only ones that nourish the malformation. These techniques are usually indicated for deep-seated angiomas or those situated in areas that are functionally important. Ligation of the carotid artery in the neck has few indications, and we have performed this on only three patients with very special angiomas, some having coexistent saccular aneurysms, after very careful angiographic studies, and we had excellent results.

Embolization procedures: no matter what material is used as an embolus, we consider this highly unsuitable. It is certain that the fistulas exercise a suction effect on the injected emboli, but in spite of this we can never be sure that the emboli will not also occlude normal vessels. Neither do we believe that the calibre of all the afferent arteries can be measured with certainty so that the size of the emboli can be accomodated to them. If not measured properly, they can cross the fistulas and produce pulmonary embolism. The use of liquid or viscous substances that become solidified at the level of



Fig. 4. Huge cerebellar AVM with enormous drainage, dependent basically of the distal part of the superior cerebellar artery. Total extirpation

the fistulas and, because of their plastity, can occlude them, are much better than the solid ones. Theoretically this should be the technique chosen, but it still is not sufficiently perfected. We have personally embolized four AVMs with a mixture of Lipiodol and bone wax $(70^{\circ}/_{0} \text{ and } 30^{\circ}/_{0} \text{ respectively})$, that should have a fusion point above 45 °C, and is injected while still melted (Brizzi's method). The handling of this mixture is technically difficult, and we regret the death of one patient because of pulmonary embolism. We have applied this technique especially in the selective embolization of the afferent branches of the external carotid artery at a dural fistula, as previously described. In the future it may be possible that embolization will be the chosen method in treating certain malformations when we can manage to use the afferent arteries to the angioma during temporary occlusion of the rest of the arterial tree, and have correct intraoperative anteriographic controls. We also think that we should study the possibility of placing an "umbrella" in the venous system returning to the heart to impede pulmonary embolisms. These techniques, in which we are personally interested, are still not perfected. In the territory of the internal carotid artery, with microsurgical techniques we can selectively occlude branches of the Sylvian artery or of the anterior cerebral system and embolize only the branches that feed the malformation.

Cryocoagulation is recommended by Walder³³, but our experience is limited to only one case, in which we had negative results as to the obliteration of the malformation, and there were serious transitory sequelae.

The field of *contraindications*, in the surgery of AVMs, is becoming more and more restricted, because of advances in operating techniques. Even if it is not a formal contraindication, the extreme bleeding capacity of the malformations in the very young and the vascular sclerosis in the elderly should be taken into account. Fortunately, the greatest number of angiomas that we have seen, in spite of being congenital, are in the second, third, and fourth decades of life, and there are few contraindications in these age groups.

Excessive extent of a malformation that can occupy a hemisphere, or its location in vital zones, such as the brain stem or an important functional zone like the parietal lobe of the dominant hemisphere, constitute formal contraindications. Experience has shown us that in all cases of doubtful indication, because of extent or of unfavourable location, we must tend towards a conservative attitude, even if the malformation has bled, especially now that have CAT at our disposal to monitor the location, extent, and evolution of a haematoma. Generally, AVM haemorrhages are less serious than those of the saccular aneurysms, and many years can elapse between two consecutive haemorrhages. Large angiomas have less tendency to bleed than the small fistulae.

Reasons		
Unfavourable location		13
Excessive size of lesion		5
Insufficient symptomatology		2
Refusal of operation		7
Death on admission		5
	Total	32

Table 8. Arteriovenous Malformations not Operated on

Table 9. Operator	y Mortality,	and Its	Causes ((146)	operations)	ļ
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Attributed to intervention	pulmonary embolism (embolized AVM of the ten anaesthetic cardiac arrest (before removal) postoperatory meningitis uncontrollable intraoperative haemorrhage (14- old patient with a temporal AVM with of carotid participation) immediate postoperative haemorrhage delayed postoperative death with periodic co ness (two angiomas of the thalamus with as haematoma, and one angioma of the tempor probable thrombosis of the Galen vein system (A the choroid tissue of the third ventricle) gastrointestinal haemorrhage	-month- external nscious- sociated al lobe) VM of	1 1 1 1 3 1 1
		Total	10 (7%)
Not attributed	operations in second and third degree comas with intraparenchymatous haematomas	(AVMs	6
		Total	6 (4%)
Mortality rate			16 (11%))

In Table 8 we show the reasons for not operating on 32 of our patients, and in Table 9 we show the operative mortality and its causes. In most neurosurgical units, as in ours, the feeling is that in the long run it is better to give surgical treatment, performed by trained specialists with adequate technical means, and that this gives better results with less mortality and morbidity than conservative attitudes. If we exclude from the sixteen patients that died, the six that were operated on in coma as a consequence of voluminous intraparenchymatous haemorrhage (and all would have undoubtedly if treated conservatively), the mortality rate of our series would be 70/0, which we consider very low, and this is the best defense of our intervention criteria.

Coexistence of Malformations

We have four examples of the coexistence of an AVM with saccular aneurysms, whose treatment has not presented any special difficulties.

Differential Diagnosis

Given the typical arteriographic characteristics of the AVM, there are rarely any diagnostic doubts. However, we have observed an angioblastic meningioma and a glioma whose feeding and drainage vascularization were absolutely like those of angiomas, causing false diagnoses, that were corrected at surgery.

Final Remarks

It is our view that, at present, cerebral arteriovenous malformations are no longer rare in most Neurosurgical Services and, thanks to the technical advantages that microsurgery offers, they are susceptible to complete extirpation in a very high percentage of cases. For this reason we propose that, to establish valid comparisons between the various statistics, these should specify the total number of cases observed in addition to the total number of cases operated on, and indicate the main reasons for choosing a conservative treatment and rejecting a surgical one. There is reason to believe that very low percentages of mortality, as have been seen in some series, may coincide with low percentages of operability. We consider it necessary to put forward in this way the percentages of mortality in relation to the malformations, since undoubtedly hemispheric and polar AVMs should not be classed with AVMs situated in medial or paramedial structures or in the posterior fossa. We also consider that malformations with a predominant participation by the external carotid artery and draining to the dural sinuses form a well-defined group which needs different therapeutic procedures from the rest of the arteriovenous malformations of the brain.

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