High-flow vascular malformations of the brain in pediatrics: Experience in a tertiary care children’s hospital

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ABSTRACT

Introduction. High-flow vascular malformations of the brain are uncommon in pediatrics. The objective of this study is to establish the differences among these pathologies and group them by age at onset, clinical manifestations, and angioarchitecture.

Population and method. This was a retrospective and observational study. The medical records, imaging studies, and procedure protocols of patients seen at Hospital J. P. Garrahan diagnosed with vascular malformations of the brain between January 2010 and January 2020 were analyzed.

Results. A total of 183 patients met the inclusion criteria. It was possible to identify 131 patients with arteriovenous malformations with a nidus (AVMs) and 52 with direct fistulas (without a nidus), including 19 vein of Galen aneurysmal malformations, 23 pial fistulas, and 10 dural fistulas. The average age of patients was 105 months for AVMs, 1.7 months for vein of Galen aneurysmal malformations, 60.5 months for pial fistulas, and 41 months for dural fistulas.

Conclusion. Based on their angioarchitecture, high-flow vascular malformations of the brain presented a nidus (AVMs) or direct fistulas (vein of Galen aneurysmal malformations, pial fistulas, and dural fistulas). AVMs were observed in early childhood, especially due to intracranial hemorrhage. Direct fistulas occurred in the first stage of life, commonly with heart failure.

Key words: central nervous system vascular malformations, dural arteriovenous fistula, pial veins, vein of Galen malformations, pediatrics.

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INTRODUCTION

Vascular malformations of the brain in pediatrics are uncommon.\(^1\) For a better understanding, they may be divided into high- and low-flow. The former include, on the one side, arteriovenous malformations with a nidus (AVMs) and, on the other side, vascular malformations with direct fistulas, i.e., without an interposed nidus. These include vein of Galen aneurysmal malformations (VGAMs), pial fistulas (PFs) and dural fistulas (DFs).
Cavernous hemangiomas, telangiectasias, and developmental venous anomalies are considered low-flow vascular malformations, and are less aggressive.2,3

High-flow vascular malformations (HFVMs) are made up of arteriovenous shunts without capillaries. They are localized in the meninges and the brain. HFVMs are the most common cause of spontaneous intracranial hemorrhage in children.4 They may lead to death, cause sequelae and brain development alterations.1 The analysis of their characteristics in a patient series may provide clinicians with a better understanding of these pathologies.

OBJECTIVE
The objective of this study is to establish the differences among HFVMs and group them by age at onset, clinical manifestations, and angioarchitecture.

METHODS AND POPULATION
This was an observational and retrospective study conducted between January 2010 and January 2020 in Hospital J. P. Garrahan. Medical records, computed tomography (CT), computed tomography angiography (CTA), magnetic resonance (MR), magnetic resonance angiography (MRA), cerebral digital subtraction angiography (cDSA), and procedure protocols from all patients younger than 18 years diagnosed with HFVMs were analyzed. Patients who missed any of the above-mentioned information were excluded.

Age groups were divided according to the World Health Organization (WHO) into newborn (NB): 0-28 days; infant: 1 month to 1 year; preschool child: 1 year to 5 years and 11 months; school-age child: 6 years to 12 years; and adolescent: 13 years to 18 years.5 To analyze the data in our series, the NB group was subdivided into newborn baby in the strict sense (NB, 0-24 hours of life) and neonate (1-28 days old).

The number of patients by pathology, age at onset, sex, clinical manifestations, angioarchitecture, and associated conditions were recorded. Results were described as absolute frequency and relative frequency, and measures of central tendency and dispersion were determined.

NBs and neonates with VGAMs were assessed using the Bicêtre score, which encompassed cardiac, hepatic, renal, and neurologic functions (Annex). The study was approved by the Institutional Ethics Committee. Data were anonymized.

RESULTS
A total of 183 cases of patients with HFVMs were collected (Table 1). Based on their angioarchitecture, they were divided into two groups: AVMs, which had a nidus interposed between arteries and veins, and direct fistulas, which did not feature a nidus. The latter were sub-divided depending on the meningeal layer involved: VGAMs (velum interpositum cistern/subarachnoid space), PFs (pia mater), and DFs (dura mater). In turn, DFs were grouped based on their association with dural sinus malformation (DSM) into type I or II. Lastly, pathologies were classified as associated with cerebrofacial arteriovenous metameric syndrome (CAMS) or with capillary malformation-arteriovenous malformation syndrome (CM-AVM).

Frequency, sex, age
AVMs were the most common HFVMs in our series and accounted for 70% (131 patients) of cases; among these, 50.6% occurred in males. They were prevalent in school age. The mean age at onset was 105 months (median: 126, range: 30-218).

VGAMs accounted for 10% (19 patients) of HFVMs and were prevalent among males (58%). They were observed in NBs and infants. The mean age at onset was 1.7 months (median: 0.03, range: 0-18).

PFs accounted for 13% (23 patients) of HFVMs and were prevalent among males (65%). They occurred at all ages, and a peak incidence was observed in preschool age. The mean age at onset was 60.5 months (median: 46, range: 0-204). Single PFs corresponded to 65% of cases, and multiple ones, to 35%.

DFs accounted for 6% (10 patients) of HFVMs. DFs associated with type I DSM were more common (3 males and 2 females), with a mean age at onset of 7 months (median: 4, range: 0-22). Two male patients aged 3 and 5 months had a DF associated with type II DSM. Three DF cases were not associated with DSM (2 males and 1 female), with a mean age at onset of 129 months (median: 132, range: 96-158) (Figure 1).

Clinical manifestations
AVMs: A total of 100 patients (76%) started with intracranial hemorrhage. Eleven
Table 1. Summary of the patient series studied at Hospital de Pediatría Juan P. Garrahan. The table shows the different groups of pathologies with the corresponding number of patients, presentation, and age at onset by case

| Diagnosis | Sub-diagnosis | Age group | Presenting symptoms |
|-----------|---------------|-----------|---------------------|
| Dural fistula | No DSM = 3 patients | School-age child = 10 patients | Hemorrhagic stroke, Strabismus |
| DSM I = 5 patients | Adolescent = 1 patient | Incidental |
| DSM II = 2 patients | Infant = 4 patients | Cardiopulmonary abnormalities, Seizures, Frontal mass, Massive epistaxis, facial vein dilation, Exophthalmos |
| Pial fistula | NV = 6 patients | Cardiopulmonary abnormalities = 5 patients, Seizures |
| Infant = 2 patients | Hemorrhagic stroke, Seizures, Asymptomatic (studied for skin spots) |
| Preschool child = 13 patients | Headache = 3 patients, Hemiparesis, Developmental delay and macrocephaly |
| School-age child = 3 patients | Seizures, Hemorrhagic stroke, Developmental delay |
| Adolescent = 4 patients | Seizures = 2 patients, Hemorrhagic stroke, Strabismus |
| VGAMs | NB = 13 patients | Cardiopulmonary abnormalities = 6 patients, Asymptomatic (antenatal diagnosis) = 6 patients, Generalized hypotonia |
| Neonate = 3 patients | Macrocephaly, Seizures, Cardiopulmonary abnormalities |
| Infant = 2 patients | Macrocephaly |
| Preschool child = 1 patient | Seizures |
| Arteriovenous malformation | Preschool child = 14 patients | Hemorrhagic stroke = 11 patients, Headache = 3 patients |
| with nidus | School-age child = 77 patients | Hemorrhagic stroke = 63 patients, Seizures = 8 patients, Progressive neurological deficit = 5 patients, Headache |
| Adolescent = 32 patients | Hemorrhagic stroke = 26 patients, Seizures = 3 patients, Progressive neurological deficit, Headache = 2 patients |

DSM: dural sinus malformation; NB: newborn; VGAMs: vein of Galen aneurysmal malformations.
patients (8%) had seizures not associated with hemorrhage. Six patients (5%) had a headache. Six patients (5%) had progressive neurological deficit. In 8 patients (6%), symptoms were not clearly described in the medical record and it was not possible to contact them.

VGAMs: Heart failure was the most common manifestation and it was observed in 7 patients. Six patients were asymptomatic and were diagnosed antenatally. Macrocephaly occurred in 3 cases; seizures, in 2; and generalized hypotonia, in 1.

PFs: The most common events were heart failure in 5 patients, and intracranial hemorrhage, in 5. Four patients had seizures. One patient had motor impairment, 1 had strabismus, and 4, a headache. Cognitive impairment was observed in 2 patients. One asymptomatic patient was diagnosed due to skin spots.

DFs: The 5 patients with DFs associated with type I DSM started with heart failure, seizures, frontal mass, epistaxis, and exophthalmos (due to orbital venous-lymphatic malformation in the setting of CAMS), respectively. The two cases associated with type II DSM were diagnosed due to palpable thrill behind the ears. The three cases that were not associated with DSM had strabismus, intracranial hemorrhage, and one corresponded to an incidental finding. Table 1 shows clinical manifestations further divided by age group.

**DIAGNOSIS**

AVMs: A CT was done during an acute event to look for bleeding. A CTA helped to establish the malformation’s angioarchitecture, i.e., feeding, nidus, and draining veins. A 4-vessel cDSA helped to establish the lesion’s angioarchitecture more precisely (Figure 2).

The brain parenchyma, the nidus topographic location and its angioarchitecture were assessed with a MR and MRA, respectively. A functional MR was used to establish the relation between the AVM and the eloquent areas.

VGAMs: With a transfontanellar ultrasound, the median prosencephalic vein (MPV) was seen as a pulsatile, rounded, median image behind the third ventricle with turbulent flow based on the Doppler test. A MR showed the brain status (brain atrophy, calcifications, and increased ventricular size). An MRA evidenced the angioarchitecture of VGAMs. A CT was done only to rule out acute hemorrhage. A cDSA was performed during embolization to prevent dye toxicity and radiation (Figure 3).

PFs: A transfontanellar ultrasound showed a hypoechogenic image with flow on the brain surface. The MR and MRA were done to see the brain parenchyma and PF angioarchitecture. A CT and CTA were used for acute diagnosis of intracranial hemorrhage. A cDSA was the most sensitive study; it was only performed during embolization in NBs and infants (Figure 4).

**Figure 1. Area chart with detailed pathology distribution by age group**
DFs: The transfontanellar ultrasound done in the setting of DFs found a hypoechoic mass in the venous sinus with flow to the walls. The MR and MRA showed a partially or totally thrombosed and enlarged sinus with arteriovenous fistulas on the walls, from branches of the external carotid artery. A CT and CTA were used if bleeding was suspected. The cDSA was the diagnostic method of choice and was done during embolization in NBs and infants (Figure 5).

**Figure 2. Arteriovenous malformations with a nidus**

A. cDSA. Left internal parietal AVM. The components of this structure are pointed out.
B. 3D cDSA reconstruction.
C. CT. Acute bleeding in deep AVM.
D. CTA. The nidus (circle) is seen clearly.
E. MRA. The nidus and the deep draining vein (arrow) are clearly seen.
F. MR: T1. The nidus (arrow) is seen.
**TREATMENT**

AVMs: Microsurgery, embolization, and radiotherapy (if the nidus was smaller than 3 cm) were done separately or combined to remove the nidus in AVMs.

VGAMs: The drugs used to treat heart failure included dobutamine and milrinone. Embolization was the procedure of choice as of 4 months old in patients with a Bicêtre score of 13 or higher. An emergency procedure was done if the Bicêtre score was between 8 and 12.

Embolization was not performed if the score was lower due to an extremely poor prognosis, even with fistula occlusion.

PFs: An intravascular treatment was the preferred option at our hospital because it is less invasive in PFs (Figure 4, G-I).

DFs: Embolization alone or in association with open surgery was done, depending on the case.

**Relation to vascular syndromes**

One case of DF associated with type I

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**Figure 3. Vein of Galen aneurysmal malformation**

Mural type
A. cDSA: artery with direct fistula into the MPV (arrow).
B. MR: T2 immediately post-embolization. The MPV shows 2 different intensities due to partial thrombosis (central hyperintensity).
Note the ventricular dilation.
C. Follow-up MR 6 months after treatment. The MPV is smaller. Clear ventricular size reduction.

Choroidal type
D. MR: T1. The circle depicts the network typical of choroidal VGAMs and the falcine sinus.
E. cDSA during embolization. The MR findings are repeated.
F. and G. MRA. The angioarchitecture (circle) is the same as that observed in the cDSA during treatment.
DSM and cerebrofacial venous metameric syndrome (CVMS) and 3 cases of AVM with CAMS were reported. One case of AVM, 3 of VGAMs, and 3 of PFs were associated with CM-AVM.

**DISCUSSION**

HFVMs in pediatrics are uncommon. They may lead to death, disabilities, and abnormal brain development in the first months of life.\(^1,6,7\) There are two well-defined HFVM groups. AVMs, which have an interposed nidus malformation between arteries and veins, and direct arteriovenous fistulas, where the nidus is absent. Although these pathologies are individually recognized in the bibliography, they have not been frequently addressed in combination.\(^8,9\) This hinders the possibility of making a statistical comparison with other series because some studies consider PFs and AVMs as a single entity (but they are not) and others also included low-flow malformations. A clear example is a statement that has been repeated in several articles that VGAMs account for 30% of vascular malformations, without specifying what type of pathologies they encompass.\(^10\)

**Figure 4**

**Left frontal pial fistula**
A. cDSA: dye injection in the left carotid artery. Frontal Sylvian branches with a fistula into a single cortical vein (arrow).

The left anterior cerebral artery is not visualized due to the middle cerebral artery.
B. MRA showing the same image. The arteries of the opposite hemisphere are seen.
C. MR: T2. Brain atrophy in the left frontal lobe. Flow gap images due to dilated cortical veins.
D. and E. MRA post-embolization. No signs of arteriovenous fistula.
F. MR post-embolization. No flow gaps observed due to venous dilations. Marked left frontal atrophy.

**Right temporal pial fistula**
G. CTA showing the venous dilation (circle) and feeding artery (arrow).
H. MR: flow gap image of the dilated vein in T1. Note the brain atrophy.
I. cDSA: the image shows the fistula receiving flow from feeding arteries, the dilated cortical vein, and the transverse sinus.
In this article, all HFVMs were addressed as a whole. AVMs are an abnormal shunting between arteries and veins through a collection of interposed tortuous abnormal vessels (nidus) without intervening capillaries. In our series, they occurred as of 3 years old, and were more common in school-age. In the bibliography, AVMs are extremely rare in NBs and infants. Intraparenchymal hemorrhage was the most common manifestation. They occurred alone or together with other pathologies, such as CAMS and CM-AVM. Such associations have already been reported by other authors.

The presence of a nidus malformation with feeding arteries and early draining veins help to diagnose this condition. Although angioarchitecture may be observed with a CTA and MRA, a cDSA is still the best method for an accurate lesion determination. The study titled A randomized trial of unruptured brain arteriovenous malformations (ARUBA) showed that patients older than 18 years had a higher risk for morbidity and mortality with treatment, compared to what was observed in patients with asymptomatic AVMs. No study has supported such conduct in asymptomatic AVMs in patients younger than 18 years. The goal of AVM treatment is to completely remove the nidus to prevent bleeding and rebleeding through microsurgery, embolization, radiotherapy or a combination of these.

VGAMs result from an abnormality in development between weeks 6 and 11 of gestation, a stage when there is normally a shunt between the choroidal arteries and the MPV in the fetus. The MPV is a precursor to the vein of Galen and the straight sinus. In VGAMs, such embryonic shunts persist and lead to MPV dilation. The latter venous structure appears larger in imaging studies. Therefore, VGAMs do not feature a properly formed vein of Galen and the pathology is actually in the MPV. They are classified into choroidal, mural, and mixed.

**Figure 5**

Patient with DF, type I DSM in the left transverse sinus, and CVMS.

A. cDSA: DF that is fed by a branch of the left internal carotid artery (arrow).
B. cDSA: DF feeding from branches of the right external carotid artery (arrow). Dilated left transverse sinus (oval).
C. MRA: the arrow points at the dilated transverse sinus.
D. MR: T1. Left orbital venous-lymphatic malformation (arrow). Dilated left transverse sinus (oval).
E. MR w/gadolinium: paraventricular cavernous hemangioma (oval). DF feeding artery (arrow). Dilated transverse sinus (double arrow).
F. MR: T1. The arrows point at the dilated venous system caused by DF draining to the sinus and, from here, to the cortical veins.
depending on the type of fistula. Choroidal VGAMs have a vascular network with multiple feeding choroidal arteries that flow into the MPV. In mural VGAMs, there is a direct shunt into the MPV. In the case of mixed VGAMs, both types of fistulas coexist.24

As in other series, VGAMs were more common in NBs,22,23 Heart failure and cerebrospinal fluid (CSF) circulation disorders due to cerebral venous hypertension leading to macrocephaly or hydrocephalus were the most frequent events in our series of VGAMs, consistent with the bibliography.8

A DF is an arteriovenous shunt without an interposed nidi observed in the dura matter. It is very uncommon, so very few cases were included in our study. DFs associated with type I DSM are congenital; morphologically speaking, they feature a dilated venous sinus with fistulas in the walls. Highly diverse clinical manifestations were observed, which prevented us from making a comparison with other series where macrocephaly and seizures were the most common events.26 DFs associated with type II DSM feature a direct fistula between the occipital artery and a stenotic sigmoid sinus. In our series, they presented with thrill, a sign already described in the bibliography.26,27

In our patients, DFs not associated with DSMs were not different from those observed in adults, and are distinct from the former. They may have been formed due to thrombosis in the dural sinus caused by a traumatic brain injury or hypercoagulable states.28 A PF is a direct shunt between arteries and veins at the level of the pia mater. They may be single or multiple.29 In our series, they were observed in NBs and also in other age groups. They are arteriovenous fistulas in the brain cortex. In general, the vein is dilated, resulting in a striking finding in imaging studies.30 Heart failure and intraparenchymal hemorrhage were the most common events. This information is consistent with other publications as well, as the higher incidence of brain hemorrhage among older children.31

In NBs and infants, the preferred option for the diagnosis of VGAMs, PFs, and DFs is a combination of ultrasound, MR, and MRA. A CT and CTA are reserved for emergencies. In this age group, a DSA should be performed during embolization to prevent excessive radiation and dye administration.6

The goal of intravascular treatment is to occlude the fistulas and correct the abnormalities caused by heart failure and cerebral venous hypertension. Three vascular syndromes were observed in our series. CM-AVM is made up of multifocal capillary malformations on the skin in the form of spots, accompanied by PFs, VGAMs or AVMs in the brain. CAMS corresponds to AVMs affecting the brain and the face in a metameric pattern resulting from the common origin of the vascular cells in these locations. CVMS is the venous metameric counterpart of CAMS; it corresponds to DFs associated with type I DSM with brain cavernous hemangioma and orbital venous-lymphatic malformation.13,33

CONCLUSIONS
Based on angioarchitecture, HFVMs were divided into two groups: AVMs (interposed nidi) and direct arteriovenous fistulas, made up of VGAMs, PFs, and DFs. AVMs occur as of 3 years old, with a peak incidence in school age. Intracranial hemorrhage and seizures were the most frequent manifestations.

VGAMs, PFs, and DFs were observed in the early stage of life. Heart failure, CSF circulation disorders, and intracranial hemorrhage were the most common events. ■

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### ANNEX

**BICÊTRE SCORE**

| Score | Cardiac function | Cerebral function | Respiratory function | Hepatic function | Renal function |
|-------|------------------|-------------------|----------------------|-----------------|---------------|
| 5     | Normal.          | Normal.           | Normal.              | -               | -             |
| 4     | Overload, no medical treatment. | Subclinical, isolated EEG abnormalities. | Tachypnea, finishes bottle. | - | - |
| 3     | Failure, stable with medical treatment. | Non-convulsive, intermittent neurologic signs. | Tachypnea, does not finish bottle. | No hepatomegaly, normal hepatic function. | Normal. |
| 2     | Failure, not stable with medical treatment. | Isolated convulsions. | Assisted ventilation, normal saturation FIO₂ < 25 %. | Hepatomegaly, normal hepatic function. | Transient anuria. |
| 1     | Ventilation necessary. | Seizures. | Assisted ventilation, normal saturation FIO₂ > 25 %. | Moderate or transient hepatic insufficiency. | Unstable diuresis with treatment. |
| 0     | Resistant to medical treatment. | Permanent neurologic signs. | Assisted ventilation, desaturation. | Abnormal coagulation, elevated enzymes. | Anuria. |

FIO₂: fraction of inspired oxygen. See text for explanation.