LUPINE PUBLISHERS

Progressing Aspects in Pediatrics and Neonatology

DOI: 10.32474/PAPN.2025.05.000204

ISSN: 2637-4722

Review Article

Vein of Galen Malformation – Experience of the Last 13 Years in a Romanian Reference Center

Maria Alexandra Fleacă*, Alexandra Bratu, Mihaela Bizubac, Veronica Marcu, Cristina Filip, Carmen Herișeanu, Carmina Nedelcu, Cătălin Cîrstoveanu

Marie-Sklodowska Curie Emergency Hospital for Children, Bucharest, Romania

***Corresponding author:** Maria Alexandra Fleacă, Marie-Sklodowska Curie Emergency Hospital for Children, Bucharest, Romania

Received: November 22, 2025 **Published:** December 09, 2025

Introduction

The vein of Galen malformations (VoGMs) are mainly correlated with the retention of an embryonic pattern of vascularity, characterised by vein of Galen dilation and formation of arteriovenous communications that give rise to risk of systemic shunting, causing cardiac dysfunction, vascular steal and venous hypertension. Diagnosis is made by colour Doppler ultrasound in the first place, followed by MRI or CT scan that can provide more details on vascular architecture, while angiography remains the gold standard for VoGMs follow-up.

In terms of therapeutic options, endovascular embolisation is considered to be the most suitable treatment strategy, but in resource-limited settings lacking curative intervention, focus should be on medical management aiming to counteract progressive heart failure and postpone endovascular intervention.

Materials and methods

We consulted the literature regarding VoGM via PubMed Central (published not earlier than 2010) and analysed concurrently our practical experience and evolving treatment strategies over the last 13 years. We present this retrospective series of 9 eloquent cases for the Galen vein malformation from NICU of Marie-Sklodowska Curie Emergency Hospital for Children in order to illustrate how this pathology has known an upward evolutionary trend in terms of diagnosis, treatment and survival rate in our unit.

The inclusion criteria in our case series were any suspicion of arteriovenous malformation notified at the level of the quadrigeminal cistern during prenatal morphological screening or any dilated, pouch-shaped structure contiguous with the sagittal sinus, displaying turbulent Doppler signal during transfontanellar ultrasound screening at admission following echocardiographic findings relevant for cardiac decompensation with diastolic steal from the aorta or unexplained cause of persistent pulmonary hypertension, irrespectively of gestational age or associated conditions.

We aimed to stratify the clinical presentation of our patients using the Bicêtre neonatal score (grading system by symptom group descending from 5 to 1 points according to severity) and systematise our diagnostic and therapeutical means over time in order to refine both our own strategies and those from other units lacking endovascular treatment perspectives.

Table 1:

Points	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 baby bottle	–	–
3	heart failure, stable with medical treatment	non-convulsive, intermittent neurologic signs	tachypnea, does not finish baby bottle	no hepatomegaly, normal hepatic function	normal

2	heart failure, not stable with medical treatment	isolated convulsion	assisted ventilation, normal saturation FiO2 < 0,3	hepatomegaly, normal hepatic function	transient oliguria/ anuria
1	necessity for invasive mechanical ventilation	seizures	assisted ventilation, normal saturation FiO2 > 0,3	moderate or transient hepatic failure	unstable diuresis with medical treatment
0	resistant to medical therapy	permanent neurological signs	assisted ventilation, repeatable desaturations	abnormal coagulation, elevated enzymes	anuria

Results and discussions

Our patients were predominantly male, full-term newborns with satisfactory adaptation to extrauterine life, half of them with antenatal diagnosis (by ultrasound or fetal MRI), and the other half most frequently diagnosed by incidental cerebral ultrasound or after auscultation of a cardiac murmur followed by heart ultrasound documenting diastolic steal from the aorta in the carotid territory towards the cerebral circulation. For some patients, the diagnosis was completed by brain MRI or angio-CT

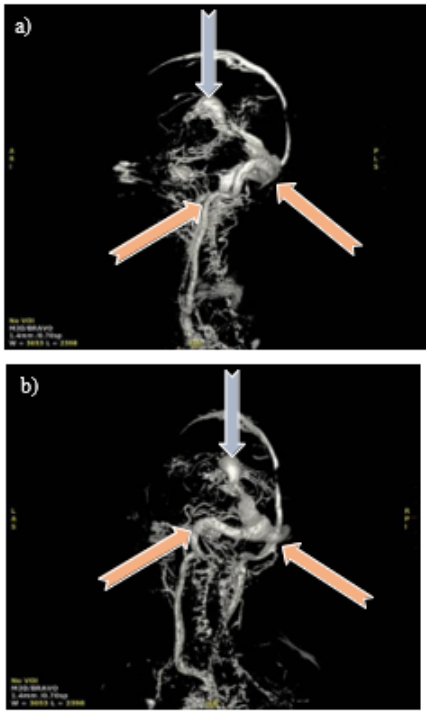

Figure 1: 3D reconstruction angio-MRI: a). *Sagittal view of the VoGM, b). Posteroanterior view in coronal plane Aneurysmal median prosecephalic vein drained by the right sinus (blue arrows). Transverse/sigmoid sinuses and internal jugular veins of increased caliber (pink arrows). Multiple vascular pathways at the level of the basal nuclei connecting the anterior and posterior circulation. Carotid, vertebral arteries and basilar trunk exerting mass effect on the bulb*.

for a better understanding of the Galen vein morphology and confirmed variable degrees of compression with secondary changes in the appearance of the midline and ventricular system. None of the patients underwent an angiographic study. 2/3 of patients recorded Bicêtre scores ranging from 4 to 7 in the framework of neuroradiological risk factors or severe heart failure refractory to medical treatment, involving progressive organ failure. The surviving patients had a Bicêtre score between 16 and 20.

Medical treatment was focused on providing inotropic support and tightly controlled peripheral and pulmonary vasodilation with the aim of overriding high output heart failure and better ensuring end-organ perfusion. All patients maintained their renal function within normal limits, spontaneously or stimulated by diuretics, not necessitating continuous renal replacement therapy in the context of cardiac decompensation. Most of the patients underwent liver failure and flow mediated pulmonary hypertension, imposing

the necessity for respiratory support in order to reduce cardiac workload. Half of the newborns expressed anomalies of the nervous system (ventriculomegaly, attenuation of the cortical-subcortical differentiation, chronic ischemia of certain vascular territories with subsequent atrophy) due to impaired cerebral hemodynamics.

We instituted targeted volume lung-protective ventilation, recruitment techniques for preventing ventilation/perfusion mismatch, but also non-invasive ventilation strategies in order to facilitate successful weaning and prevent bronchopulmonary dysplasia. We ensured inhaled nitric oxide therapy to offset pulmonary hypertension and maintained a careful balance between permissive ventilatory hyper/hypocapnia.

We practiced point-of-care ultrasound (assessing tissue Dopplers, diastolic markers, LV strain, flow patterns), NIRS and aEEG monitoring. In the framework of systemic hypoperfusion due to blood flow diverted from abdominal aorta, we propose efficient renal replacement therapies if renal impairment occurs and early extracorporeal membrane oxygenation support before multiple organ failure sets in.

Conclusions

Compared to early days of VoGM diagnosis in Romania, when cases were often identified postnatally and significant systemic decompensation was already present, patients are now increasingly diagnosed antenatally and admitted to our NICU for supportive care until the optimal timing for curative intervention.

However, the elevated morbimortality of this condition in our country enunciates a sad reality and raises the issue of prenatal underdiagnosis of this pathology with vital risk for newborns, even though progress has been made in terms of fetal screening and treatment.

In the context of endovascular treatment unavailability in our unit, we acknowledge the importance of assessing the clinical impact of the VoGM at the time of diagnosis and providing customised intensive care in order to maintain the cardiorespiratory and hemodynamic stability until transfer is feasible in centers with expertise for endovascular embolisation.

DOI: 10.32474/PAPN.2025.05.000204