

Research Article

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MANAGEMENT OF THE VEIN OF GALEN ANEURYSMAL MALFORMATION CASES PRESENTING IN THE NEONATAL PERIOD



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Abstract

Objective: Vein of Galen aneurysmal malformations (VGAM) are rare congenital vascular anomalies with challenging management, especially in neonates with severe hemodynamic compromise. This study evaluated outcomes of neonatal VGAM treated with endovascular embolization at a single center.

Materials and Methods: We retrospectively analysed 14 patients diagnosed antenatally or symptomatic within 28 days, managed between 2005 and 2025. Data included demographics, clinical and cardiac status, imaging, Bicêtre scores, treatment, and outcomes. Per protocol, embolization followed cardiopulmonary stabilization and could occur beyond the neonatal period.

Results: Of the 14 neonates, 12 (85.7%) underwent embolization, while two (14.3%) died before intervention due to poor general condition. The mean gestational age was 38.8±1.0 weeks, and the mean birth weight was 3,283±292 g. The majority of the cases (66.7%) were classified as mixed-type VGAM. The mean Bicêtre score was 11.0±3.9. Embolization was performed at a mean of 176±149 postnatal days, requiring an average of 2.1 sessions per patient. Pulmonary hypertension and cardiomegaly were the most frequent cardiac findings (75% and 58.3%, respectively). The embolic agents included were n-butyl cyanoacrylate (NBCA) and ethylene vinyl alcohol copolymer (EVA). The mortality rate among embolized patients was 33.3%. Survivors exhibited normal neurodevelopment in 75% of the cases.

Conclusion: Endovascular embolization is a viable strategy for VGAM cases presenting in the neonatal period, although mortality remains substantial in patients with severe cardiac compromise. Early diagnosis, multidisciplinary management, and long-term follow-up are essential for optimizing survival and neurological outcomes.

Keywords Vein of galen malformation • Neonate • Endovascular treatment • Hydrocephalus • Heart failure • Bicêtre score



INTRODUCTION

Vein of Galen aneurysmal malformations (VGAM) are rare, complex congenital vascular anomalies resulting from abnormal arteriovenous shunts between cerebral arteries and the median prosencephalic vein during early embryogenesis (1). Although infrequent, VGAM constitutes about 30% of pediatric intracranial vascular malformations and can lead to life-threatening conditions, especially in neonates (2). Advances in prenatal imaging and postnatal neuroimaging techniques have improved early diagnosis, allowing better perinatal management strategies. Historically associated with poor outcomes, VGAM treatment has significantly evolved with the development of endovascular embolization techniques, leading to decreased morbidity and mortality compared to traditional surgical approaches (3-5).

The clinical presentation of VGAM varies depending on the patient's age. In neonates, high-output cardiac failure is the predominant manifestation due to massive arteriovenous shunting. In contrast, older infants and children typically present with hydrocephalus, seizures, or neurological deficits, whereas adolescents and adults may experience headaches or hemorrhagic events. Early diagnosis and intervention are crucial, as severe heart failure and multiple organ dysfunctions are common in untreated or late-treated neonates (6-8).

This study aims to present our single-center experience in the management of neonatal VGAM cases, emphasizing endovascular treatment outcomes, and showcasing representative clinical examples. By reflecting on the therapeutic challenges and patient outcomes, we aim to contribute insights to the existing literature and underline the importance of timely diagnosis and multidisciplinary management in improving the prognosis for affected neonates.

MATERIALS AND METHODS

Study design and patient selection

This retrospective study included neonates diagnosed with VGAM and treated at Istanbul University, Istanbul Faculty of Medicine, between 2005 and 2025. The study was conducted following approval from the Institutional Clinical Research Ethics Committee (Date: 09.02.2024, No: 03). Cases were enrolled in the study if antenatal diagnosis or clinical presentation occurred during the neonatal period (≤ 28 days). A total of 14 neonates diagnosed with VGAM were identified from the Neurosurgery and Neuroradiology departmental archives. Two of these patients died before endovascular treatment could be performed due to rapid clinical deterioration. Since no follow-up data were available for these cases, they were excluded from the statistical analyses, which were performed only among patients who survived long enough to receive treatment or clinical follow-up. However, these early fatal cases were still described qualitatively to reflect the full clinical spectrum of the disease. This

study represents a continuation and expansion of our previously reported single-center VGAM series published in 2017, incorporating additional patients managed at our institution with extended follow-up (9).

Inclusion and exclusion criteria

Patients were included in the study if they had an imaging-confirmed diagnosis of VGAM and either an antenatal diagnosis or clinical symptom onset within the first 28 postnatal days. Cases were excluded if available data were insufficient to allow reliable extraction of the clinical course, treatment details, or outcomes.

Data collection

Demographic data (gestational age, birth weight, gender), prenatal diagnostic information, clinical presentation, echocardiographic findings, VGAM subtype classification (mixed, choroidal, mural), Bicêtre clinical score, and timing and number of embolization sessions were recorded. Detailed procedural data, including embolic agents used and post-procedural outcomes, were also collected.

Preoperative evaluation

Patients underwent comprehensive neurological examination and cardiac evaluation with echocardiography to assess heart failure severity, pulmonary hypertension, and other associated cardiac anomalies. Imaging studies included cranial ultrasound, magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), and digital subtraction angiography (DSA) for diagnosis and procedural planning.

Treatment protocol

Endovascular embolization was the primary treatment strategy. Procedures were performed under general anesthesia using a trans-arterial femoral approach in most cases. Embolic materials included n-butyl cyanoacrylate (NBCA) and ethylene vinyl alcohol copolymer (EVA, Onyx). In selected cases, coil embolization was combined with liquid embolic agents to achieve a more effective occlusion of high-flow feeders and enhance procedural control. The choice of embolic agent and the number of sessions were determined based on angiographic findings and the patient's clinical condition. The primary therapeutic goal was a staged and controlled reduction of arteriovenous shunting while preserving physiological venous drainage and minimizing the risk of venous hypertension or ischemia. Consistent with our institutional protocol for fragile hemodynamics, interventions were submitted until cardiopulmonary stabilization when feasible. Consequently, the timing of the first embolization often fell beyond the neonatal period, despite neonatal presentation.

Hydrocephalus associated with VGAM was managed according to the center's standardized protocol. Cerebrospinal fluid (CSF) diversion



was delayed until after endovascular embolization, as an early shunting can worsen venous outflow obstruction and intracranial hypertension. Patients were monitored with serial cranial ultrasound or MRI following each embolization session. A ventriculoperitoneal (VP) shunt was considered only in cases of persistent or progressive ventriculomegaly accompanied by clinical signs of raised intracranial pressure after hemodynamic stabilization. The typical waiting period before shunt placement ranged from two to six weeks post-embolization, depending on the resolution of venous congestion and ventricular size. Endoscopic third ventriculostomy (ETV) was considered when favorable anatomy was permitted, though most cases required VP shunting due to venous distortion of the third ventricular floor.

Outcome measures

Primary outcomes included survival, neurodevelopmental status,

and the need for secondary interventions such as VP shunt placement. Complications related to the procedure and clinical follow-up outcomes were also analyzed. To contextualize timing, we report the age at first embolization separately from age at presentation, emphasizing that all cases met neonatal presentation criteria.

RESULTS

A total of 14 neonates diagnosed with VGAM were included in the study. Among them, 12 patients (85.7%) underwent endovascular embolization, whereas two patients (14.3%) were deemed unsuitable for intervention due to poor general condition and died within the first two weeks postnatally. All 14 cases were diagnosed antenatally or became symptomatic within the neonatal period (≤ 28 days). By protocol, definitive embolization was performed after stabilization, yielding a mean age at first embolization of 176 days (Table 1).

Table 1. Clinical and procedural characteristics of neonates with Vein of Galen aneurysmal malformations treated by endovascular embolisation

Patient no	GA (weeks)	Birth weight (g)	Gender	Diagnosis week	Cardiac findings	VGAM type	Bicêtre score	Postnatal age at procedure	No. of procedures	Embolized arteries	Embolic agent	Outcome	VP shunt
1	38	3150	M	34	HF, PHT	Mixed	12	Day 3	1	Choroidal	NBCA	Discharged, no sequelae	No
2	38	3220	F	37	HF, PHT	Mixed	11	Day 2, 1 mo, 1 yr	3	Choroidal	NBCA	Death	No
3	37+4/7	3290	M	35	PHT, PFO, TR, CM	Mixed	9	Day 3	1	Right ACA	NBCA + EVA	Discharged, no sequelae	No
4	41+1/7	3205	F	37	PHT, RVD/RAD, R-L shunt, Ao arch, VSD, CM	Choroidal	12	Day 2, Day 9, 4 yrs	3	Right ACA, Left PCA, PMCA, PLCA	NBCA	Discharged, no sequelae	No
5	38	3800	M	26	PHT, TR, CM, RVD/RAD	Mixed	11	Day 18, 27	2	Right ACA, PCA, PLCA	NBCA	Death	No
6	38	3500	M	26	PHT, CM, RVD/RAD, TR	Mixed	8	Day 1	1	Right ACA	NBCA	Death	No
7	39+3/7	2770	M	38	PHT, CM, RVD/RAD, TR, PDA	Mixed	12	Day 2, 4, 8	3	Left PCA, Left ACA	EVA	Discharged, no sequelae	No
8	39	3250	M	39	PHT, CM, PFO, TR	Mixed	12	Day 25, 40, 9, 5 mo	3	Left PCA, Left ACA	EVA	Severe neurological impairment	Yes
9	39+4/7	3015	M	37	PHT, CM, RVD/RAD, TR	Mixed	12	Day 3	1	Left ACA, Left TMPA	NBCA	Death	No
10	39	3300	M	38	HF	Choroidal	14	Day 8, 19, 11 mo, 2 yrs	4	Ant. choroidal, ACA	Coils + NBCA	Discharged, no sequelae	No
11	39	3250	M	37	None initially	Choroidal	13	Day 14	1	Ant. & post. choroidal	Coils + NBCA	Discharged, no sequelae	Yes
12	38+2/7	3200	M	38	HF, PDA	Choroidal	7	Day 3	2	Left ACA, post. choroidal	Coils	Severe neurological impairment	No

GA: Gestational age, M: male, F: Female, HF: Heart failure, PHT: Pulmonary hypertension, PFO: Patent foramen ovale, TR: Tricuspid regurgitation, CM: Cardiomegaly, RVD/RAD: Right ventricular/atrial dilation; Ao arch: Aortic arch anomaly, VSD: Ventricular septal defect, VSD: Ventricular septal defect, PDA: Patent ductus arteriosus, VGAM: Vein of Galen aneurysmal malformation, HT: Hypotension, ACA: Anterior cerebral artery, PCA: Posterior cerebral artery, TMPA: Thalamoperforating artery, PMCA: Posteromedial choroidal artery, PLCA: Posterolateral choroidal artery, NBCA: N-butyl cyanoacrylate, EVA: Ethylene vinyl alcohol copolymer, VP: ventriculoperitoneal.



The cohort included 11 males (78.6%) and 3 females (21.4%). The mean gestational age at birth was 38.8 ± 1.0 weeks, and the mean birth weight was $3,283 \pm 292$ grams. The mean week of prenatal diagnosis was 34.6 weeks (range: 26–39 weeks). Most patients (66.7%) were diagnosed with the mixed type of VGAM, while 4 patients (33.3%) had the choroidal type.

The mean Bictre clinical score at presentation was 11.0 ± 3.9 (range, 6–17), with lower initial Bictre scores observed among non-survivors compared with survivors (mean 7.5 vs. 12.5, respectively). However, the sample size did not allow for formal statistical testing. The mean postnatal day at first embolization was 176 ± 149 days, and the average number of embolization procedures per patient was 2.1. The most commonly embolized arteries were the anterior choroidal artery, posterior cerebral artery, and posterior choroidal artery. Regarding embolic materials, NBCA was used in 58.3% of cases, EVA in 16.7%, and a combination of NBCA and EVA in 25%. In addition, coils were used with NBCA in two patients and as a standalone embolic material in one patient, particularly for high-flow lesions, to facilitate flow reduction and improve procedural control. The mean clinical follow-up duration was 42 months (range: 8–96 months).

Among the patients who underwent embolization, four (33.3%) died despite intervention, all presenting of severe heart failure. Of the eight surviving patients, six (75%) exhibited normal neurodevelopmental outcomes during follow-up, while two patients developed hydrocephalus requiring VP shunt placement.

Cardiac evaluations revealed that pulmonary hypertension and cardiomegaly were the most common findings, present in 75% and 58.3% of cases, respectively. Tricuspid regurgitation was noted in 50% of patients. Other anomalies, such as patent foramen ovale, ventricular septal defects, and right-to-left shunts, were less frequently observed. Clinical and procedural characteristics of all neonates are summarized in Table 1.

Case presentations

Case 1

A male neonate born via cesarean section at 39 weeks of gestation was admitted to the Neonatal Intensive Care Unit following prenatal detection of a VGAM at 38 weeks. Initial cardiological evaluation revealed signs of heart failure, and the patient was started on intravenous furosemide (2×0.5 mg/kg/day). Transfontanel ultrasonography (TFUS) confirmed VGAM, and subsequent cranial MRI, MRA, and magnetic resonance venography demonstrated a 29×21 mm aneurysmal dilatation at the Galen vein level. There was associated dilation of the straight sinus, torcular herophili, transverse sinus, and occipital sinus.

Magnetic resonance angiography revealed large arteriovenous fistulous connections from bilateral posterior choroidal arteries, along with contributions from anterior cerebral,

pericallosal, and thalamoperforators arteries. These findings were consistent with a choroidal-type VGAM according to the Lasjaunias classification, characterized by multiple bilateral choroidal feeders.

Bictre scoring was used to monitor the clinical course. On the third postnatal day, the patient experienced focal seizures and was started on phenobarbital; the Bictre score was calculated as 14. Due to clinical deterioration and increased oxygen demand by the eighth day, the patient was referred for endovascular intervention.

Under general anesthesia, endovascular embolization was performed via right femoral artery access. Angiography confirmed high-flow arteriovenous shunting from the right anterior choroidal and anterior cerebral arteries. Embolization was performed using coils and NBCA. Initial control angiography showed approximately 20% occlusion. A second embolization was performed on postnatal day 19, targeting an additional AV fistula. Third and fourth embolization sessions were performed at 11 months and 2 years of age, respectively, achieving near-complete occlusion of the lesion.

The patient was neurologically stable after all procedures, did not require CSF diversion, and was discharged without complications. Follow-up imaging confirmed successful embolization, and clinical assessments showed normal neurodevelopment (Figure 1).

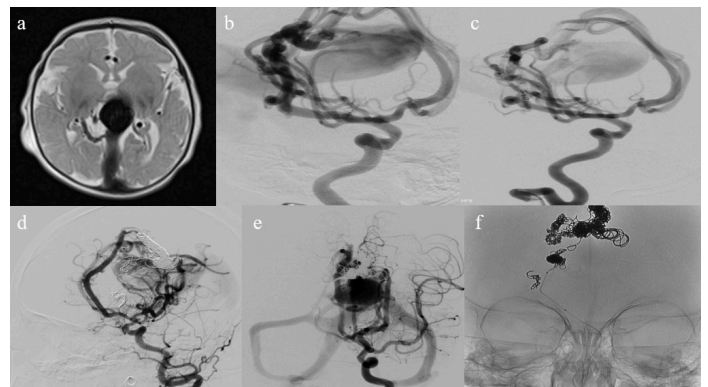


Figure 1. Imaging findings of the first neonatal Vein of Galen aneurysmal malformation (VGAM) case. a. Axial T2-weighted cranial MRI showing a markedly dilated vein of Galen consistent with VGAM. b-c. Pre-embolization digital subtraction angiography (DSA) in anteroposterior and lateral projections demonstrating a high-flow arteriovenous malformation with major feeders from the anterior choroidal artery and pericallosal arteries. d. Post-embolization DSA revealing partial occlusion of the main feeders after the first intervention. e. Subsequent DSA showing further embolization success with decreased vascularity. f. Postoperative skull X-ray illustrating embolization material distribution after multiple embolization sessions.

Case 2

A male neonate was diagnosed antenatally with a VGAM at 37 weeks of gestation during routine ultrasonographic follow-up. He was delivered via elective cesarean section at 39 weeks of gestation. At birth, the initial physical and neurological examination was normal. The patient was admitted to the Neonatal Intensive Care Unit (NICU)

for postnatal monitoring. Cardiac evaluation revealed no initial abnormalities.

During the first postnatal week, progressive macrocephaly was noted, with a head circumference measuring 44 cm (>99th percentile). Transfontanelle ultrasound revealed marked dilation of the ventricular system and an arteriovenous malformation with turbulent high-velocity flow localized at the Galen vein level. Electroencephalography (EEG) performed during the same period was within normal limits.

Repeat TFUS at two weeks of age demonstrated progression of both the malformation and ventriculomegaly. Cranial computed tomography (CT) showed a large midline lesion (measuring approximately 45.7×51.2×48.7 mm), consistent with VGAM and associated hydrocephalus. Diagnostic cerebral angiography confirmed bilateral anterior and posterior choroidal artery feeders, with the VGAM nidus measuring 84.8×55.3 mm. The diagnosis was consistent with choroidal-type VGAM.

During the same session, endovascular embolization was performed. Both internal carotid and vertebral arteries were catheterized, and the major arterial feeders were selectively coiled and embolized with NBCA (Figure 2). Postoperatively, the patient was stable and transferred to the NICU for monitoring. Despite embolization, macrocephaly progressed (head circumference 47.5 cm, >99th percentile), prompting neurosurgical intervention. A VP shunt was placed. Neurological examination post-shunt showed no significant abnormalities.

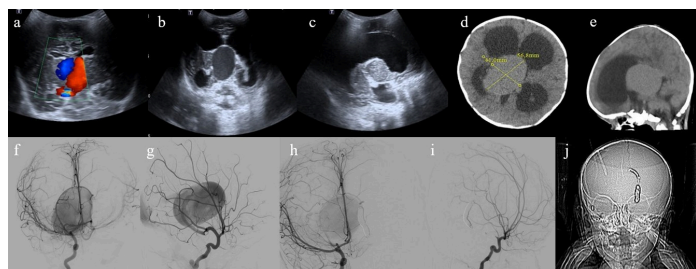


Figure 2. Imaging findings of a newborn male diagnosed antenatally with Vein of Galen aneurysmal malformation (VGAM) at 37 weeks of gestation and delivered via cesarean section at 39 weeks. a-c. Postnatal transfontanelle ultrasound images showing ventriculomegaly and a large arteriovenous malformation at the Galen vein level with high-velocity flow on Doppler imaging. d-e. Cranial computed tomography (CT) scans demonstrating severe hydrocephalus and a markedly dilated aneurysmal sac measuring approximately 84.8×55.3 mm. f-g. Pre-embolization digital subtraction angiography (DSA) images in anteroposterior and lateral views, revealing anterior and posterior choroidal arteries as the main feeders. h-i. Post-embolization DSA images showing successful occlusion of the main feeding arteries after coiling and embolization. j. Post-procedural cranial radiograph illustrating the distribution of embolization material.

Follow-up MRI and MRA demonstrated significant thrombosis of the malformation, with minimal residual flow near the previously embolized posterior choroidal feeders. Ventricular size was reduced. The patient remained asymptomatic and neurologically intact during

regular follow-up visits. Developmental assessments throughout infancy and early childhood revealed age-appropriate milestones with no evidence of neurological deficit.

DISCUSSION

Vein of Galen aneurysmal malformation is a rare, high-flow congenital vascular anomaly resulting from the persistence of the embryonic median prosencephalic vein, leading to complex arteriovenous shunting. It often presents in the neonatal period with high-output cardiac failure, multiorgan dysfunction, and occasionally hydrocephalus. Despite significant advances in prenatal imaging, diagnosis, and treatment techniques, VGAM continues to pose major therapeutic challenges (10, 11). Although our cohort is described as neonatal in presentation, definitive endovascular treatment was frequently scheduled beyond the neonatal window. This reflects a staged strategy prioritizing cardiopulmonary stabilization and controlled flow reduction rather than urgent complete occlusion in fragile neonates. The observed mean of 176 postnatal days for the first embolization is therefore a management choice, not a contradiction of neonatal presentation.

The mean interval of approximately six months between diagnosis and the first embolization reflects a deliberate, protocol-based management strategy rather than a delay in care. In neonates presenting with severe cardiac failure and high-output physiology, initial medical stabilization is crucial to reduce peri-procedural risk. Our institutional protocol prioritizes achieving hemodynamic and cardiopulmonary stability before proceeding with definitive endovascular intervention. In addition, technical factors such as vessel caliber, venous drainage maturation, and anesthetic safety often necessitate postponing embolization beyond the neonatal period. Therefore, the timing observed in this series represents a clinical necessity and safety-driven decision, constituting one of the most critical aspects of our management approach.

In our series, the majority of patients were male, diagnosed in the late third trimester, and predominantly exhibited mixed-type VGAM, consistent with previously reported data (4). Prenatal diagnosis allowed for early stabilization and multidisciplinary management planning, although survival outcomes remained primarily influenced by the severity of cardiac failure rather than by the timing of diagnosis alone (12). Pulmonary hypertension and cardiomegaly, detected in a substantial proportion of our cohort, were strongly associated with poor prognosis, as emphasized by Fullerton et al. (13).

Our experience supports the now widely accepted approach favoring staged endovascular embolization. Instead of attempting complete obliteration in a single session, which may precipitate catastrophic venous hypertension, gradual reduction of arteriovenous shunting allows better adaptation of fragile neonatal hemodynamics (14-16).

Regarding embolic materials, both NBCA and EVA were utilized. NBCA, a fast-polymerizing adhesive agent, enables rapid vessel occlusion but requires highly precise injection to avoid non-target embolization. By contrast, EVA has a slower polymerization profile and non-adhesive properties, allowing for more controlled and prolonged embolization, especially useful in complex or tortuous vascular anatomies. Lesion flow dynamics, arterial anatomy, and procedural goals guided the choice between the materials (17-20).

Hydrocephalus was observed in several patients, reflecting its multifactorial nature in VGAM. Venous hypertension, which causes impaired CSF absorption, mechanical aqueductal obstruction by the dilated Galen vein, and intrinsic cerebral venous congestion, all contribute (21). In our cohort, VP shunting was required after embolization in select cases. As emphasized in previous reports, delaying CSF diversion until after hemodynamic stabilization is crucial to avoid worsening venous outflow obstruction. Although ETV is a potential alternative, anatomical constraints caused by massive VGAMs may limit its feasibility (22, 23).

The Bicêtre score proved valuable for evaluating patients' clinical stability and guiding intervention timing; however, relying solely on this scoring system may be insufficient. It lacks detailed integration of angiographic architecture, venous drainage patterns, and cerebral perfusion dynamics, which are essential for comprehensive risk stratification. Newer proposals advocate for the inclusion of such imaging-based parameters alongside clinical scoring (24). In our cohort, a clear tendency was observed for lower Bicêtre scores among non-survivors, supporting its established prognostic utility in neonatal VGAM. While the small sample precluded formal statistical correlation, this pattern aligns with prior reports identifying Bicêtre score thresholds <8-10 as predictive of unfavorable outcomes and limited treatment tolerance. Therefore, our findings reinforce the value of early functional assessment in guiding treatment decisions and parental counseling.

In our study, the neonatal mortality rate among embolized patients was 33.3%, consistent with the literature for neonates with severe heart failure (25). Survivors exhibited favorable neurodevelopmental outcomes in 75% of cases, emphasizing that early, carefully timed embolization can lead not only to survival but also to preserved neurological function. However, two patients developed hydrocephalus requiring permanent CSF diversion, one of whom exhibited significant neurological sequelae, underlining the importance of vigilant long-term follow-up.

Overall, VGAM management demands a multidisciplinary approach, involving pediatric neurology, neurosurgery, neuroradiology, neonatology, and intensive care specialists. Institutional experience and technical expertise remain pivotal to achieving optimal results. Future directions should focus on refining classification systems, integrating functional imaging parameters, and expanding

multicenter collaborative studies to establish standardized management protocols.

This study has several limitations. First, its retrospective design inherently carries the risk of selection and information bias. Second, the relatively small sample size reflects the rarity of VGAM, especially in the neonatal population, but it may limit the generalizability of our findings. Third, the study was conducted at a single center with specialized endovascular expertise, which may not reflect outcomes at institutions with different resource availability or levels of experience. Additionally, long-term neurodevelopmental follow-up was limited for some patients, restricting the ability to fully assess cognitive and behavioral outcomes over time. Future prospective multicenter studies with standardized evaluation protocols are necessary to validate our findings and optimize management strategies for neonatal VGAM.

CONCLUSION

Vein of Galen aneurysmal malformation remains a life-threatening condition in neonates, particularly those presenting with severe cardiac compromise. Advances in endovascular embolization techniques have significantly improved survival and neurodevelopmental outcomes compared to historical data. Our single-center experience highlights that early diagnosis, careful clinical evaluation, including but not limited to Bicêtre scoring, and staged endovascular intervention are crucial to optimizing patient outcomes. Hydrocephalus remains a frequent complication, necessitating individualized CSF management strategies. Despite these advances, VGAM management continues to demand a multidisciplinary approach and specialized expertise. Future efforts should aim to refine classification systems, integrate imaging biomarkers into risk stratification models, and establish standardized, multicenter treatment protocols. Ongoing long-term follow-up is essential to monitor cognitive, behavioral, and neurological development in survivors, ultimately aiming to improve survival and quality of life.





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Peer Review	Externally peer-reviewed.
Author Contributions	Conception/Design of Study- S.N., E.K.,M.B., S.S., B.G.,A.A. Data Acquisition- S.N., M.B. Data Analysis/ Interpretation- S.N., C.I.G., D.S.,N.R.,E.K.,M.B.,S.S., B.G.,A.. Drafting Manuscript- C.I.G., D.S.,N.R.; Critical Revision of Manuscript- .M.B., S.S.,B.G.,A.A; Final Approval and Accountability- S.N., C.I.G.,D.Ş.,, N.R., E.K., M.B., S.S., B.G., A.A..
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