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# A Combination Of Vein Of Galen Aneurysmal Malformation And Bovine Aortic Arch In Newborn: A Case Report

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## ABSTRACT

Vein of Galen aneurysmal malformation is a rare intracranial arteriovenous malformation, coexistence with aortic arch anomalies is even more uncommon, carry high risk of morbidity and mortality related to endovascular procedure and poor neurological outcome. Vein of Galen aneurysmal malformation is a dilated venous pouch, a persistence of the embryonic median prosencephalic vein of Markowski. Often presents with high output cardiac failure, severe pulmonary hypertension, and systemic steal leading to hemodynamic compromise and multi organ failure. Bicêtre score scale determines the potential treatment option and prognosis of the disease. Aim of the medical treatment is initial stabilization of life threatening condition of severe heart failure followed by the definitive treatment with endovascular embolization. Treatment is a big challenge, specially if coexistence with aortic arch anomalies which increases the morbidity and mortality. We describe a term neonate found to have vein of Galen aneurysmal malformation associated with bovine aortic arch.

**Keywords:** *arteriovenous malformation; bovine aortic arch; congenital anomalies; vein of Galen malformation.*

## INTRODUCTION

Vein of Galen results from the persistence of the embryonic median prosencephalic vein (MPV) of Markowski, which is normally replaced by the internal cerebral veins during neural vascularization.[1] Dilatation of the vein due to cerebral arteriovenous malformation leads to Vein of Galen aneurysmal malformation (VGAM).[1,2] It accounts for 1% of pediatric intracranial AV malformations and occurs in fewer than 1 in 25,000 births.[1,2] At birth, it may present with high-output cardiac failure, pulmonary hypertension, and systemic steal, causing hemodynamic compromise.[2-4] VGAM with anomalous aortic arch is rare and associated with higher mortality and poor outcomes.[5-7] Bovine aortic arch, the most common anomaly, has a prevalence of 7–25%.[6-7] We report a term neonate with coexisting VGAM and bovine aortic arch.

## CASE REPORT

A term female was born via elective cesarean section to a 30 years old primigravida following an uncomplicated pregnancy with Apgar score 7/10 and 9/10, birth weight of 2900 grams. Her antenatal ultrasound finding were normal, but she did not have fetal echocardiography. Soon after birth, she developed respiratory distress and became cyanosed needing supplemental oxygen via continuous positive airway pressure (CPAP) support. Congenital cardiac anomaly was suspected in view of cardiac murmur. Arterial blood gas, complete blood count, electrolytes and inflammatory marker were within normal range. Chest X-ray showed mild cardiomegaly. Echocardiography of heart showed patent ductus arteriosus (PDA) and foramen ovale (PFO) both bidirectional shunt, severe tricuspid valve regurgitation (TR), TR pressure gradient; 50 mmHg, suggestive of severe pulmonary hypertension (PAH). In addition there was abnormal branching pattern of aortic arch with presence of bovine aortic arch (Figure 1).

There was diastolic flow reversal seen in aorta with increased blood flow in superior vena cava (SVC). Further CT angiography of heart confirmed left side type A bovine aortic arch (BAA). The patient was started on sildenafil for severe PAH, maintenance

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intravenous (IV) fluid along with IV antibiotics.



**Figure 1. Suprasternal view of echocardiography showing common origin of brachiocephalic trunk and left common carotid artery (Bovine aortic arch).**

Later IV milrinone infusion was added. The presence of retrograde flow in descending aorta gave a clue for low resistance cerebral arteriovenous malformation leading to the diastolic steal phenomenon. Auscultation of anterior fontanel had bruit which suggested possible intracranial AV malformation. Bedside head ultrasound showed dilated intracranial vessels with increased vascularity and blood flow (Figure 2).

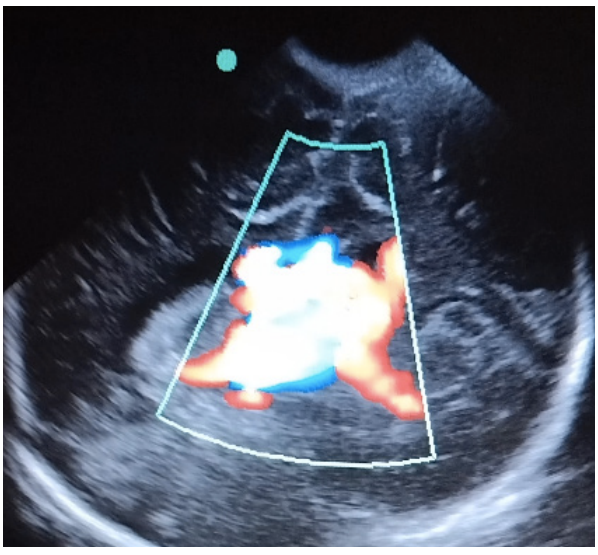


**Figure 2. Cranium ultrasound showing dilated intracranial vessels with increased flow.**

Contrast-enhanced CT of head confirmed the diagnosis of vein of Galen aneurysmal malformation, (Figure 3).Her Bicêtre score was calculated to be between 12 and 14. Neurology consultation was done who advised to continue medical management and to monitor Bicêtre score closely and planned for the endovascular embolization if Bicêtre score became less than 12 on close follow-up or at 6 months of life.

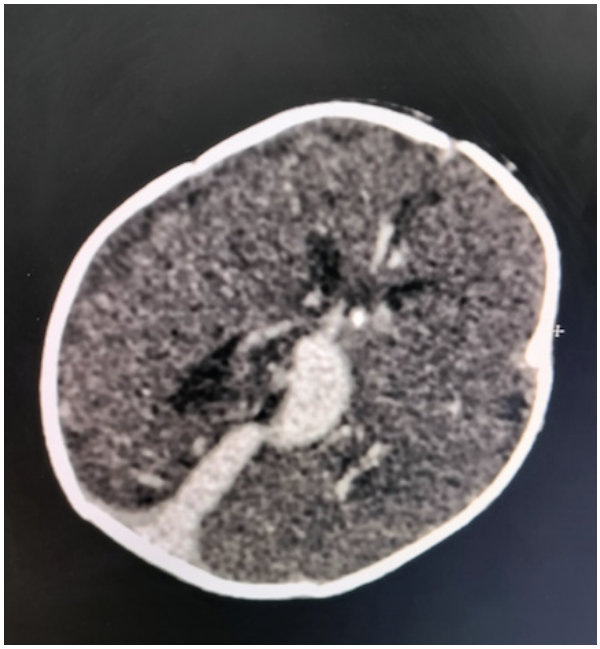


**Figure 3. Contrast enhanced CT of head showing vein of Galen aneurysmal dilations with multiple dilated feeding arteries.**

On 12$^{th}$ day of life patient developed congestive cardiac failure, so continued on CPAP & IV Lasix infusion was started. Deranged liver enzymes and coagulation profile were managed with vitamin K and plasma. Despite counseling on poor prognosis, the infant was discharged on day 18 and expired at home on day 49.

## DISCUSSION

Persistence of the embryonic MPV of markowski, which is the first vein to drain the choroid plexuses of the lateral and third ventricles between the 7$^{th}$ and 12$^{th}$ weeks of gestation and replaced by the internal cerebral veins, except most caudal portion of MPV which form the vein of Galen.[1-3] VGAM consists of multiple arteriovenous shunt draining into a dilated vein of Galen.[1] The most common feeders are the posterior choriodal arteries followed by anterior cerebral artery.[1,2] There are two types of VGAM.[4] The choroidal type, is a primitive condition, most predominant and encountered in neonatal period with severe form of cardiorespiratory symptoms. Similar to the present case, VGAM was the choroidal type and symptoms started soon after birth. In contrast to that, the mural type, direct arteriovenous fistula within the wall of the MPV, often is better tolerated and encountered in infancy with intracranial hemorrhage, headache,

seizure, neurological deficits and hydrocephalus.[1-3] Ultrasound and MRI are the essential tools for diagnosis of VGAM in utero, prognostic evaluation and optimal management of such pregnancy.[2] During fetal life VGAM well tolerated due to a low resistance placental circulation. However, after birth the flow through the VGAM increases significantly leading to high output cardiac failure, pulmonary hypertension, neurological, hepatic and renal dysfunction.[1-3] similar to the present case. Upto 80% of cardiac out put is directed toward the VGAM, which leads to increased venous return to the right side of the heart leading to heart failure and persistence of right to left shunt through PFO and PDA and subsequent PAH. In addition, circulatory steal to the VGAM produce the characteristic diastolic flow reversal in the descending aorta and potentially multi organ failure.[1-4] We have similar finding in our case except for renal failure and neurological dysfunction.

Use of Bicêtre score scale based on cardiac, respiratory, cerebral, renal and liver function to determine potential treatment option.[4] Our patient had Bicêtre score in between 12 and 14, medical management was initiated. Severe heart failure refractory to medical management patient must under go emergency endovascular embolization.[2,8,9] Currently, stage trans-arterial embolization of VGAM is the treatment of choice with favorable outcome,[2-4,8] and prevention of severe brain damage.[2] Despite treatment, the primary morbidity is hydrocephalus which leads to intellectual disability and high mortality up to 63% and also needs CSF shunting.[2,4,9] Present case had VGAM coexist with BAA, an abnormal branching pattern of the aortic arch, a left carotid artery arises from brachiocephalic trunk instead of left aortic wall.[6] Type A BAA has the common origin of left carotid artery and brachiocephalic trunk, whereas type B BAA, the left carotid artery originates directly from brachiocephalic trunk.[6] Patient with this anomaly increase risk of morbidity like infarction, hemorrhage, and vascular injury related to endovascular and surgical procedure.[5,6,7] A multicenter study showed BAA is the independent risk factor for poor neurological outcomes and has direct impact on mortality.[6,7,10] The coexistence of BAA and VGAM represented an unfavorable variation with poor outcome; prenatal screening could aid early diagnosis and prevention.

## CONCLUSIONS

VGAM is a rare intracranial AV malformation and coexistence with aortic arch anomalies is even more uncommon. The coexistence of these two condition carries high risk of morbidity and mortality related to endovascular procedure, and poor neurological outcome. This condition must be ruled out prenatally with improvement in antenatal fetal anomaly screening and fetal echocardiography. Initial treatment is to relieve the congestive heart failure, circulatory stabilization until the definitive treatment with endovascular procedure is done. Despite treatment, VGAM coexistence with other cardiac anomalies carry worse prognosis.

**Acknowledgement:** None

**Conflict of Interest:** None

**Consent:** JNMA Case Report Consent Form was signed by the patient & is attached with the patient's chart.

## REFERENCES

1. Kang HS. Update on the Vein of Galen Aneurysmal Malformation : Disease Concept and Genetics. J Korean Neurosurg Soc. 2024 May;67(3):308-14. [PubMed | Full Text | DOI]
2. Buratti S, Mallamaci M, Tuo G, Severino M, Tortora D, Parodi C, et al. Vein of Galen aneurysmal malformation in newborns: a retrospective study to describe a paradigm of treatment and identify risk factors of adverse outcome in a referral center. Front Pediatr. 2023 Jul 20;11:1193738. [PubMed | Full Text | DOI]
3. Hoang S, Choudhri O, Edwards M, Guzman R. Vein of Galen malformation. Neurosurg Focus. 2009 Nov;27(5):E8. [PubMed | Full Text | DOI]
4. Lasjaunias PL, Chng SM, Sachet M, Alvarez H, Rodesch G, Garcia-Monaco R. The management of vein of Galen aneurysmal malformations. Neurosurgery. 2006 Nov;59(5 Suppl 3):S184-94; discussion S3-13. [PubMed | Full Text | DOI]
5. Simpson RK Jr, Fischer DK, Haber LM, Mawad ME, Rose JE. Coexistence of multiple arteriovenous malformations and an anomalous aortic arch. Clin Neurol Neurosurg. 1988;90(2):169-73. [PubMed]
6. Murray A, Meguid EA. Anatomical variation in the branching pattern of the aortic arch: a literature review. Ir J Med Sci. 2023 Aug;192(4):1807-1817. [PubMed | Full Text | DOI]
7. Rotundu A, Nedelcu AH, Tepordei RT, Moraru MC, Chiran DA, Oancea A, Ma-taleru A, Costache AD, Chirica C, Grosu C, Mitu F, Leon MM. Medical-Surgical Implications of Branching Variation of Human Aortic Arch Known as Bovine Aortic Arch (BAA). J Pers Med. 2024 Jun 24;14(7):678. [PubMed | Full Text | DOI]
8. Hansen D, Kan PT, Reddy GD, Mohan AC, Jea A, Lam S. Pediatric knowledge update: Approach to the management of vein of Galen aneurysmal malformations in neonates. Surg Neurol Int. 2016 May 13;7(Suppl 12):S317-21. [PubMed | Full Text]
9. Clarke NE, Shekhawat J, Popat H, Lord DJE, Abdel-Latif ME. Vein of Galen Aneurysmal Malformation: A Case Report. Healthcare (Basel). 2024 Mar 25;12(7):716. [PubMed | Full Text | DOI]
10. Dumfarth J, Plaikner M, Krapf C, Bonaros N, Semsroth S, Rizzo JA, Fang H, Grimm M, Elefteriades JA, Schachner T. Bovine aortic arch: predictor of entry site and risk factor for neurologic injury in acute type a dissection. Ann Thorac Surg. 2014 Oct;98(4):1339-46. [PubMed | Full Text | DOI]

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