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Case Report

## Prenatal diagnosis of vein of Galen malformation; antenatal ultrasound aiding prognosis and postnatal management

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### ABSTRACT

We present a case of Vein of Galen Malformation (VGAM) diagnosed prenatally with 3 dimensional color and power Doppler ultrasonography. On the basis of prenatal imaging, the arterial feeders, drainage and secondary damage like hydrocephalus and cerebral thinning were assessed. This can be useful in counseling patients and providing a realistic view of outcome and management after birth. This is of utmost importance in a country like India, where there is disparity in services and infrastructure in maternity hospitals.

**Keywords:** Prenatal diagnosis, VGAM, Color and 3D power Doppler, Prenatal imaging

### INTRODUCTION

Vein of Galen aneurysmal malformations are rare intracranial malformations. The actual incidence is unknown but the prevalence is 1% of all neonatal intracranial vascular malformations.<sup>1,2</sup> The fetus may present with a spectrum of manifestations like nonimmune hydrops, hydrocephalus, and intracranial hemorrhage. Prenatal diagnosis of this rare arterio-venous fistula is suspected when intracerebral hypoechoic cyst is seen, which demonstrates a high velocity, low resistance blood flow. This is one of the few conditions, where use of Doppler is critical for diagnosis. Different techniques, such as conventional sonography, color Doppler imaging (CD), magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), three-dimensional (3D) sonography, and 3D color power angiography (CPA), have been described as useful diagnostic tools for a detailed examination.

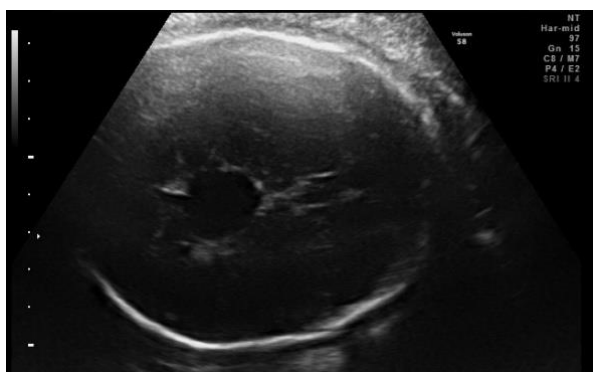
Few cases of prenatal diagnosis of VGAM have already been reported and published. The following report highlights the importance of prenatal delineation of angio-architecture of VGAM, and the role of antenatal

detection in prognostication & counselling, timing & mode of delivery. A multidisciplinary perinatal approach can help in planning further management.

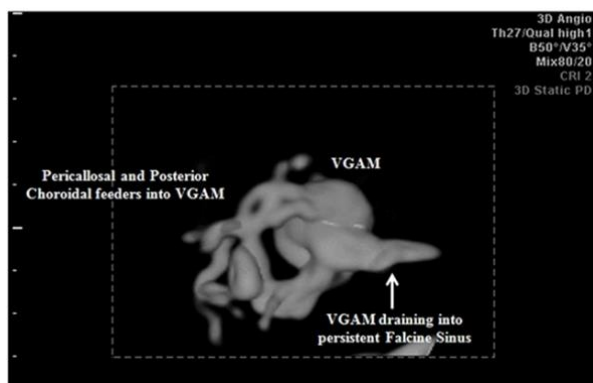
### CASE REPORT

A 31 year old healthy primigravida was referred to our unit for fetal evaluation of a brain cyst at 29 weeks gestation. The course of pregnancy was uneventful before referral. Conventional ultrasound showed a well defined, anechoic intracranial midline cystic lesion measuring 23x16 mm just above the level of thalamus (Figure 1). Colour Doppler imaging revealed high velocity, low resistance bidirectional flow in the cyst. An aneurysm of vein of Galen was diagnosed. A 3D Power Doppler Angiogram helped to depict the course of feeders and drainage of the VGAM. The VGAM drained into a dilated persistent Falcine sinus, and the straight sinus was absent (Figure 2). The rest of the brain morphology was normal, and there was no ventriculomegaly or cerebral thinning. The heart was anatomically normal, and there was no evidence of hyperdynamic circulation or hydrops. The patient did not consent for an antenatal MRI. The pediatric cardiologist and interventional radiologist were

involved. Parents were counseled that with a stable cardio-vascular status and absence of intracranial pressure signs, the prognosis is likely to be good. They were explained that the baby shall be re-evaluated at birth, and the interventional radiologist would then take a call as to whether intervention was required or not, depending on the clinical presentation and progress of VGAM. The size of the VGAM remained constant on serial ultrasounds, and the fetus also remained hemodynamically stable. At 36 weeks, a cesarean section was performed for fetal growth restriction and impaired umbilical artery Doppler. A girl weighing 2300 grams was born with an Apgar score of 9 and 9 at 1 and 5 minutes respectively.



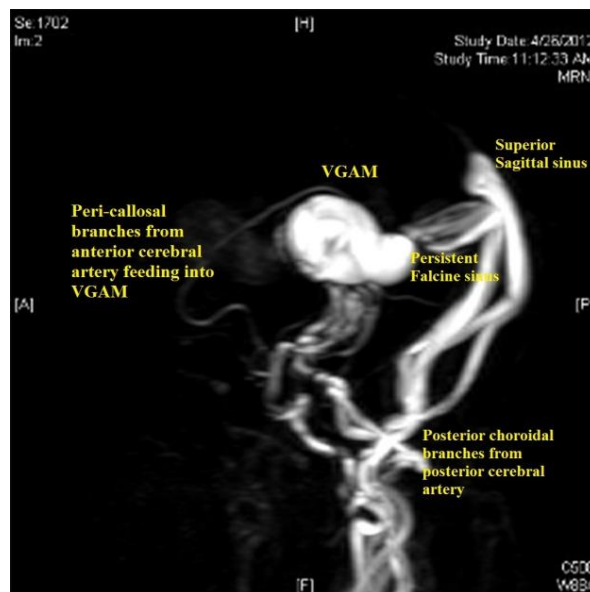
**Figure 1: Antenatal VGAM on gray-scale imaging.**



**Figure 2: Antenatal 3-D power Doppler.**

The physical examination at birth was unremarkable. Echocardiography showed a normal cardiac function with mild tricuspid regurgitation and a normal sized heart. MR angiogram confirmed the antenatal findings of feeders and drainage of VGAM (Figure 3). Interdisciplinary consultation with the neonatologists, pediatric neurologists, and interventional radiologists confirmed that the prognosis will depend upon the follow up of VGAM size and cardiac status.

Presently, the baby has not needed intervention and is neurologically normal at one year of age. The size of VGAM has regressed, and she continues to be under follow up. Need for intervention now seems less likely.



**Figure 3: Postnatal MR venogram.**

## DISCUSSION

### *Why is prenatal diagnosis important?*

The low systemic resistance of the fetus in utero can decrease the flow through the malformation and prevent cardiac decompensation. During and immediately after delivery, there is a sudden increase in systemic vascular resistance which results in an increase in blood flow through the VGAM.<sup>3</sup> This can present as an unexplained refractory congestive cardiac failure in a neonate. Hence, VGAM can cause severe morbidity and mortality in neonates. However, early detection, careful prenatal follow-up and a holistic approach can lead to a favorable outcome.

### *What is already known about VGAM?*

- Antenatal detection of intracranial cystic spaces demands color Doppler evaluation for a specific diagnosis.
- Prenatal diagnosis of VGAM is feasible.
- Size of VGAM is one of the most important prognostic factors.

### *What this report adds?*

- Use of CD and 3D CPA can help in delineating the feeders and drainage (angio-architecture) of VGAM antenatally.
- Perinatal management can be aided by angio-architecture of an antenatally diagnosed VGAM.
- In spite of the large size of VGAM, good prognosis can be anticipated in light of the fact that there were only two feeders into the VGAM, and there were no secondary effects (hydrocephalus, cerebral thinning).

- All VGAMs don't necessarily require intervention, they can be monitored by VGAM and cardiac evaluation.

The prognosis of neonates with VGAM has markedly improved. A healthy infant with normal neurodevelopmental and cardiovascular status is now a reality. This has been supported by antenatal ultrasound, which has reached a level where it can demarcate feeders, drainage and secondary effects of the VGAM, comparable to postnatal MR angiogram. This can immensely help in counseling, planning antenatal surveillance, timing, place of delivery, need for in-utero transfer and perinatal multidisciplinary care.

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