

# Approach to the management of vein of Galen aneurysmal malformation Pre and postnatal period using Bicetres scoring and Lasjaunias treatment algorithm

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## Introduction

- Vein of Galen aneurysmal malformation is a rare congenital anomaly with prevalence of 1 in 25000 pregnancies. The defect develops in the early first trimester, but the aneurysm becomes sonographically apparent just in the third trimester. In 90% of cases, there is high-output heart failure with secondary hydrops.
- Vein of Galen Aneurysm malformation is one of the most difficult intracranial vascular lesions to manage. The VOGAM is mainly divided into mural and choroidal types
- Before the advent of endovascular interventions, mortality in neonatal VGAM was nearly 90%. With the introduction of embolization techniques and advanced neonatal critical care for severe cardiopulmonary illness, mortality has decreased to around 50%. Immediate multidisciplinary neonatal assessment and care are clearly imperative

### AIMS and OBJECTIVES

Vein Of Galen malformation, which presents generally in 3 trimester requires evaluation based on Fetal Neurosonogram, Fetal echo and MRI assessment .

Though Vein of Galen Aneurysm malformation is one of the most difficult intracranial vascular lesions to manage, prenatal multidisciplinary counselling is crucial to discuss the prognosis and postnatal management

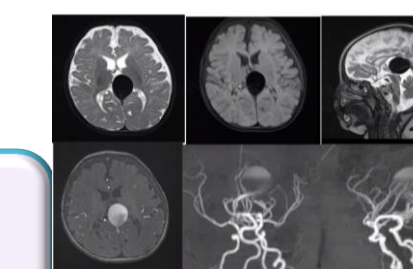
Herewith we present a case report of a fetal Vein of Galen malformation and approach to its pre and postnatal evaluation and management., using Bicetres scoring and Lasjaunias treatment algorithm

A, 31 years old, Primi spontaneous conception She was screen negative for aneuploidies in the first trimester and had a normal anomaly scan at 20 weeks. During a scan at 30 weeks' gestation she was found to have a, well defined elongated supratentorial, midline- anechoic supratentorial midline cystic structure with significant vascularity with arteriovenous flows seen within. **S/O Vein of Gallen malformation**-with no other intracranial structural abnormality, no e/o any intracranial hemorrhages. Multiple small communicating vessels are seen in Circle of Willis , flows are distorted.

**There was also mild cardiomegaly (CT ratio 0.5) and no other features cardiac failure normal growth. Serial follow up with Neurosonography and echocardiography done Multidisciplinary counselling was done involving Paed Neurologist, Cardiologist ,, Fetal medicine , Obstetrician, Neonatologist and Interventional Radiologist The requirement to deliver at tertiary care center with multidisciplinary facilities , postnatal assessment , requirement for interventional procedures and their pros and cons has been explained in detail**

The score appears >16 , the cardiomegaly settled in postnatal period and Medical management is continued until around 4 months of age when their larger baby size decreases the risks of extended embolization. Postnatal Trans arterial embolization, in a staged fashion, for partial occlusion of the VGAM is the only current treatment that has been shown to result in a safe reduction of cardiac failure

**Staged partial embolization of vein of Galen malformation was done at 5 th month and procedure is uneventful and condition of baby stable with normal neurodevelopmental reflexes for the age**



## CONCLUSION

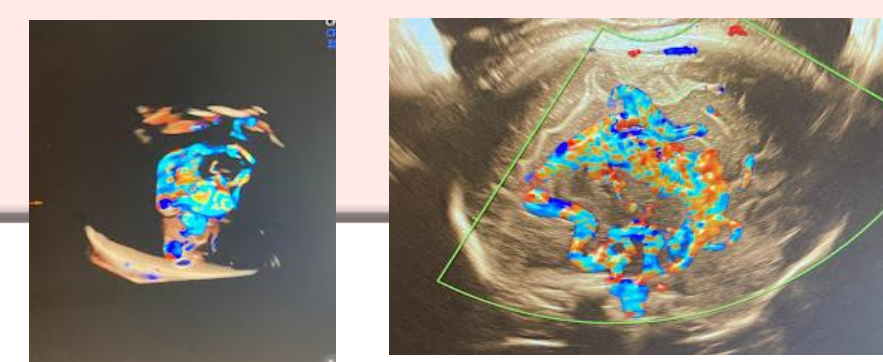
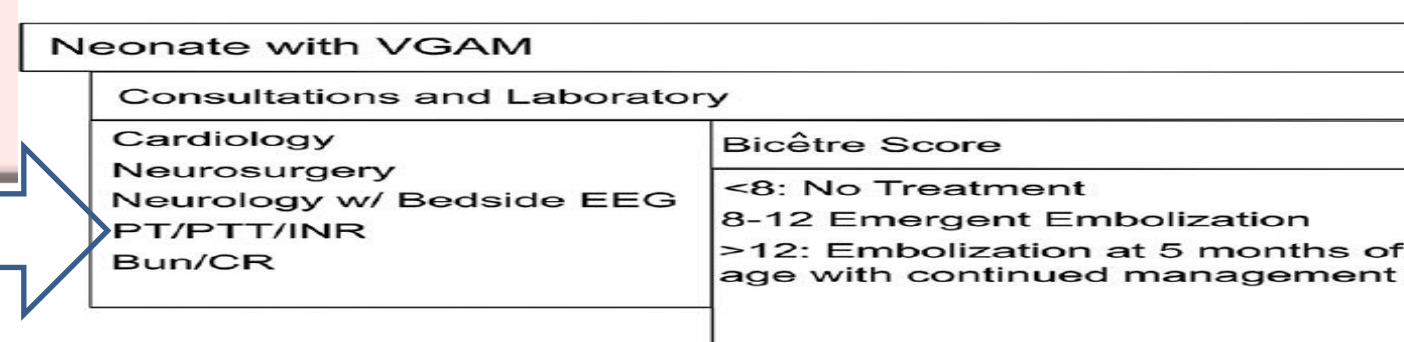
The management of VGAM is evolving, Assessment , using Bicetres scoring and Lasjaunias treatment algorithm, helps us evaluate and allow the couple choose options,

and the addition of transarterial embolization techniques is allowing increased treatment of these complex lesions with improving outcomes. For case with VGAM, Neonatology, Paediatric Neurosurgery, Paediatric Neurology, Neurointerventional endovascular, and Pediatric Cardiology services all combine to provide a tailored and individualized response to this challenging problem.

Clinical status at discharge



### Lasjaunias Treatment algorithm



### Bicetre scoring

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 bottle	-	-
3	Failure, stable with medical treatment	Nonconvulsive intermittent neurologic signs	Tachypnea does not finish bottle	No hepatomegaly, normal hepatic function	Normal
2	Failure, not stable with medical treatment	Isolated convulsion	Assisted ventilation, normal saturation FiO <sub>2</sub> <25%	Hepatomegaly, normal hepatic function	Transient anuria
1	Ventilation necessary	Seizures	Assisted ventilation, normal saturation FiO <sub>2</sub> >25%	Moderate or transient hepatic insufficiency	Unstable diuresis with treatment
0	Resistant to medical therapy	Permanent neurological signs	Assisted ventilation, desaturation	Abnormal coagulation, elevated enzymes	Anuria

\*EEG: Electroencephalogram, \*FiO<sub>2</sub>: Fractional inspired oxygen; Maximal score: 5 (cardiac) + 5 (cerebral) + 5 (respiratory) + 3 (hepatic) + 3 (renal)=21

Serial follow up with Fetal echo and Fetal NSG done , no signs of cardiac failure seen and no gross increase in size of VOG or no collaterals seen No signs and symptoms of any cerebral atrophy or dilatation of collateral seen ,Cardiopulmonary status is to be assessed in postnatal period , Immediate multidisciplinary neonatal assessment is clearly imperative Delivery planned by caesarean section at 38 weeks at centre with multidisciplinary facilities and postnatal evaluation of the newborn with VGAM is based on the Bicetre score to determine potential treatment options. (This 21-point scale gives points for the severity of signs and symptoms pertaining to the cardiac, pulmonary, neurological, hepatic, and renal systems)

## References

**Pediatric knowledge update: Approach to the management of vein of Galen aneurysmal malformations in neonates**

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