

● Case report

THE PROGNOSIS FOR FETUSES WITH VEIN OF GALEN MALFORMATION AND SYMPTOMS OF HEART FAILURE



Authors:
Agnieszka Nawara-Baran¹

1. Medical Center Ujastek- Pathology of Pregnancy Department

Prenat Cardio 2017 Jan; 7(1):83-87
DOI: 10.1515/pcard-2017-00012

Abstract

Vein of Galen Malformation (VGM) it is the most common cerebral arteriovenous malformation in fetuses and children. Usually VGM causes volume overload of the heart and can induce mass effect in the brain, causing progressive neurological impairment. Modern treatment allow on earlier therapy (before the 5th month of life). This gives the newborn with isolated VGM and heart failure a chance of survival.

This work presents case of isolated vein of Galen malformation with diagnosed cardiomegaly, monophasic flow through the tricuspid valve, large right heart, pericardiac effusion, dilatation of SVC and abnormal doppler exam. Postnatal management included aggressive medical treatment of cardiac failure and early transarterial embolization of the selected vessels feeding the aneurysm at 6 days of age. The newborn is 6 month old, growing normally without neurological deficits or developmental retardation and waiting of next stage therapy.

New techniques of treatment give fetuses with VGM and prenatal signs of cardiac insufficiency a chance of survival and healthy life, in a group of selected cases. The analysis of the cardiovascular system in fetuses with VGM according to uniform criteria is essential in order to assess the influence of these malformations on the survivability of newborns.

Key words: vein of Galen malformation, endovascular embolization; outcome.

INTRODUCTION

Vein of Galen Malformation occurs rarely and accounts for 1% of intracranial vascular abnormalities. It is the most common cerebral arteriovenous malformation in fetuses and children¹. The malformation is caused by the presence of arteriovenous fistulas directing blood flow toward the dilated, persistent proximal median prosencephalic vein (MProSV). This abnormal flow prevents involution of the embryonic vein and subsequent development of the vein of Galen. Usually VGM causes volume overload of the heart and can induce mass effect in the brain, causing progressive neurological impairment². Clinical symptoms depend on the size of malformation. According to Deloison et al. MVG occurs with other pathologies of the brain in 81% because image diagnosis is very important for proper recognition³. Prenatal diagnosis is usually carried out during the third trimester. The malformation is visible as a hypoechogenic structure located in the midline of the brain, with turbulent flow in Color doppler ultrasound. We can also use 3-D color power angiography (3-D CPA) to assess these pathological vessels of the brain⁴. Fetal magnetic resonance imaging (MRI) is important to detect associated brain abnormalities,

How to cite this article:
Nawara-Baran A.
The prognosis for fetuses with Vein of Galen Malformation and symptoms of heart failure.
Prenat Cardio 2017 Jan; 7(1):83-87

* ischemic areas and association topographic relationships between pathologically changed vessels. This exam is often a better diagnostic tool in the third trimester of pregnancy in comparison to ultrasound exam¹. Generally the outcome for fetuses with VGM is poor. Table 1 shows the factors, which worsen the outcome.

Cerebral US and MRI abnormalities	Cardiac abnormalities
Ventriculomegaly	Cardiomegaly
Hydrocephaly	Tricuspid regurgitation
Polymicrogyria	Pericardial effusion
Cortical thickening	Vena cava dilatation
Porencephaly	Hydrops
Schizencephaly	
Periventricular leukomalacia	
Intracerebral hemorrhage	

Table 1. Antenatal ultrasound (US) and magnetic resonance imaging (MRI) findings in fetuses with vein of Galen aneurysmal malformation and poor outcome according to Deloison et al.

Corresponding author: anawarabaran@gmail.com

Submitted: 2017-06-22; accepted: 2017-07-01

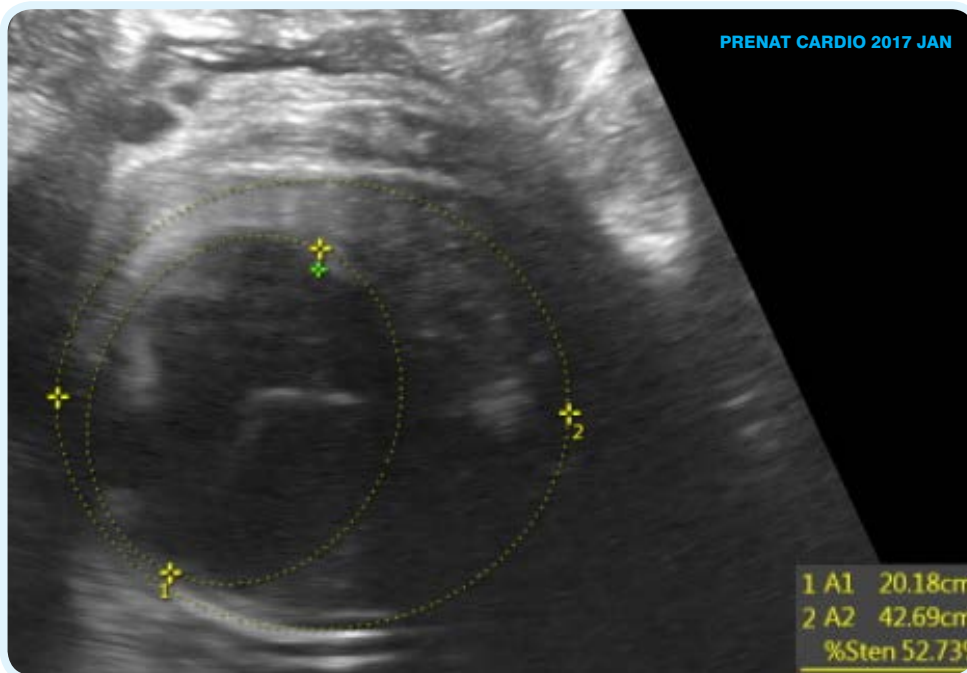


Fig. 1. Cardiomegaly

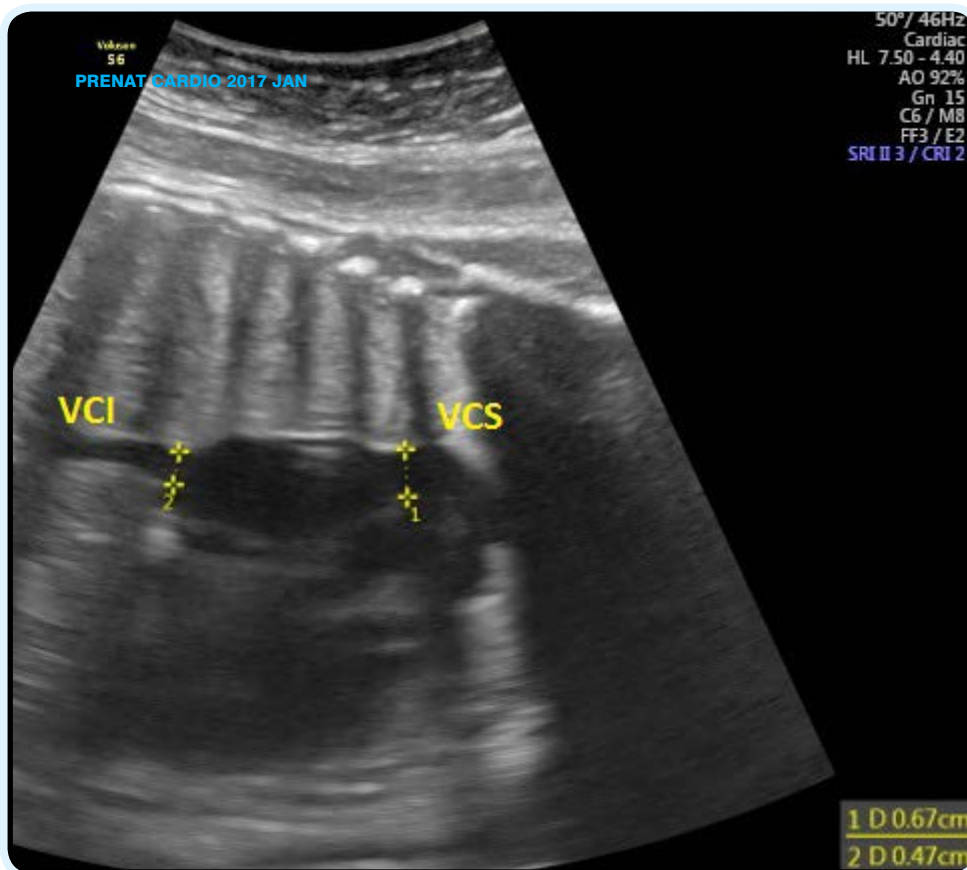


Fig. 2. Dilatation of supra vena cava

treatment in the early days of life gives the newborn with VGM and heart failure a chance to survive. There was an increase in the number of infants after treatment with good neurological development from 32% to 74%^{3,5,6}. In 2006 Lesjanias proposed a strict evaluation protocol for assessment of neonatal status before treatment, which included cerebral, cardiac, respiratory, hepatic, renal function assessment. He also changed the treatment technique by using arterial rather than venous access to endovascular therapy of VGM⁵. We can find more and more cases of fetuses with VGM and prenatal and postnatal signs of circulatory failure, witch (who) had good treatment effect in PubMed^{7,8}.

CASE REPORT

Patient gravida-2, para-1, 39+5 weeks of gestation, 24 year old, with recognized Vein of Galen malformation was admitted to the hospital for better perinatal care. The ultrasound exam confirmed the diagnosis. The fetus had: VGM (dimention:18x37mm), cardiomegaly (CTR=0,47), monofasic flow through the tricuspid valve, large right heart, pericardial effusion, dilatation of SVC (7mm), increased presser in DV (DV-PVIV=1,47), decreased MCA pulsatility index (PI=1,24), increased UA pulsatility index (PI=1,4), CPR=0,9. The pregnancy was biologically mature. The efficiency of the fetal circulation system was assessed at 7 according to cardiovascular profile score. In this situation it was decided to give birth by Caesarean section⁹. A male

2929g weight, Ap-10 was born in good condition. The newborn was stabile to the 4 day of life (heart ratio: 140-160 beat/min, blood saturation 92-100%, IBP measurement 44-52mmHg), in open incubator and closely monitored vital function. In diagnostic imaging VGM and abnormal flow through cerebral, visceral, renal arteries were confirmed.

The most important factors which worsen prognosis are cerebral defects or signs of cardiac dysfunction detected prenatally. These were the reason for termination of pregnancy in 43% cases in the Deloison et al. study in years 1999 - 2010³. In recent years, the development of treatment techniques of newborns with VGM caused a decrease in mortality from 30% to 10%^{3,5}. Starting

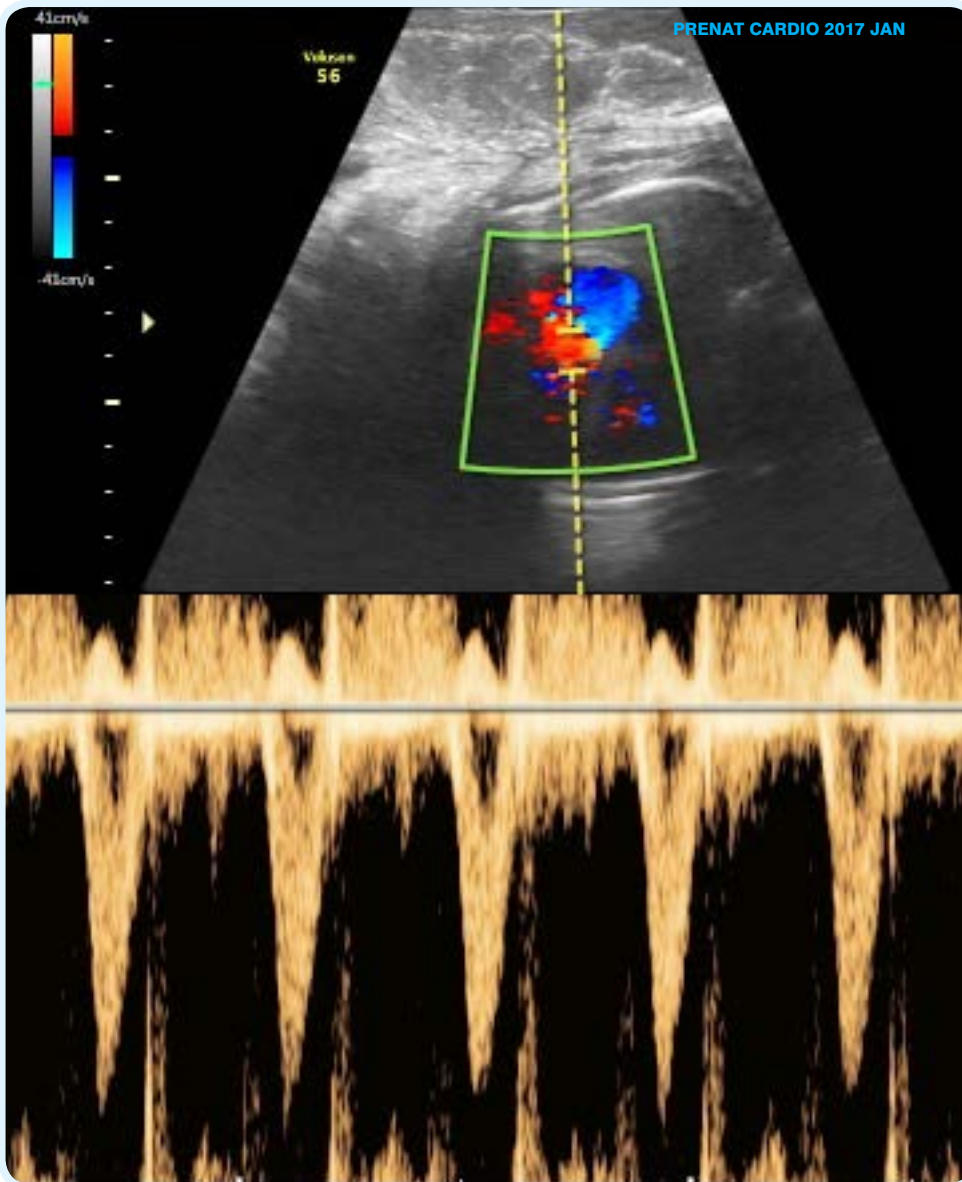


Fig. 3. Monophasic tricuspid valve flow

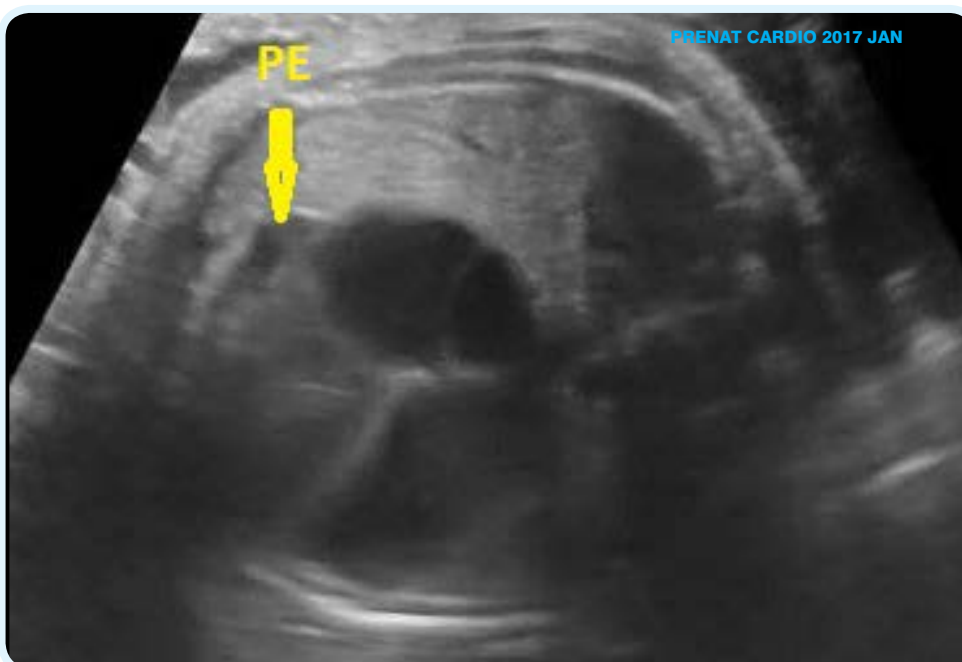


Fig. 4. Pericardial effusion

Cardiological consultation revealed the volume and pressure overload and signs of pulmonary hypertension. In the 4th day of life there was an exacerbation of the condition with an increase of heart failure and respiratory insufficiency. There was an application of Corotrope in the intravenous infusion, passive oxygen therapy, and then nCAPAP. In the 5th day the newborn was transferred to the Pathology and Intensive Therapy of Newborn Department at the Children’s Memorial Health Institute in Warsaw. There the neuroradiology team at the Neurosurgery Clinic conducted urgent first stage treatment. The newborn was treated with intravenous embolism of arteriovenous malformation which resulted in an improvement of the newborn’s condition. The next stage of therapy was performed 2 months later also with a good effect.

DISCUSSION

Modern VGM treatment begun by Lasjaunias involves the change of access to the malformation from venous to arterial. Various applications were entered endovascularly (coils, adhesive substances) which cause the embolism of the shunts supporting the malformation from the arterial side, without blocking the outflow. Earlier techniques involved venous access and the embolism caused blockage of the outflow from the malformation and increased the risk of thrombosis and hemorrhage. As a result of the gradual closing of the false track, the proper track of venous flow from this area is opened. Modern angiographs and the development of treatment allow on earlier therapy (before the 5th month of life). This gives the newborn with isolated VGM and heart

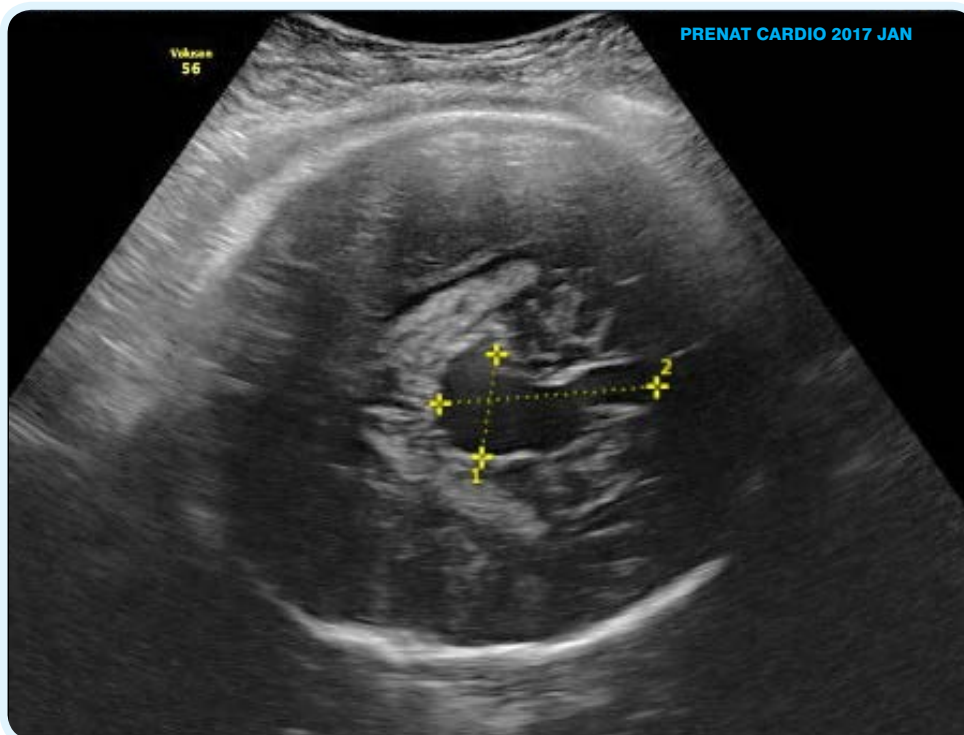


Fig. 5. Vgm-2d

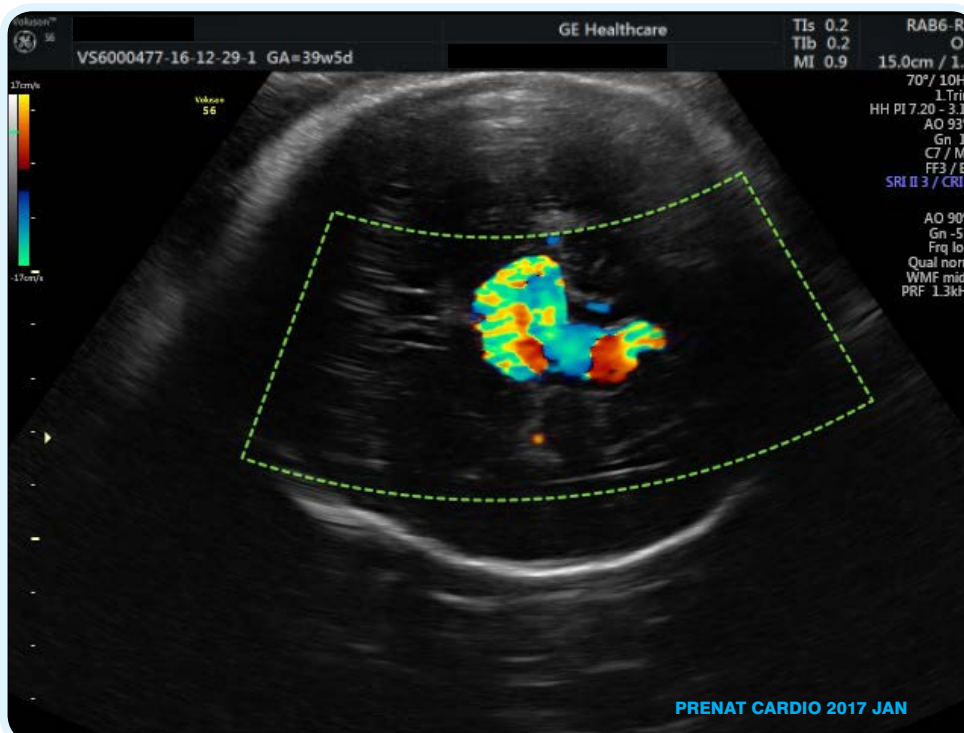


Fig. 6. Vein of galen malformation-cd

failure a chance of survival. Of course patient selection and timing remain key in the management of this condition. In the presented case the isolated VMG was recognized prenatally but the fetuses had moderate heart failure which suggested early cardiac insufficiency in the newborn and poor outcome. In 2009, in a similar case, no center in Poland would conduct treatment of a newborn with isolated VGM which in the 2nd day of life demonstrated symptoms of cardiovascular failure and the newborn died in the 7th day of life. In the presented case earlier

endovascular treatment caused the symptoms of cardiac insufficiency to subside and allowed the baby to be discharged from the hospital. This is a breakthrough in the treatment of VGM in Poland and there is hope that the cardiac insufficiency in the fetus with VGM does not have to be a death sentence for the newborn, which is confirmed by the PubMed data from recent years. Echocardiographic monitoring of fetuses with VGM with the assessment of cardiac sufficiency seems to be especially significant in this situation. In the analyzed publications the cardiac insufficiency was not evaluated in an objective way, only the symptoms were listed. Godfrey et al., after analyzing the cases from 1999-2015 only made a generally conclusion that the enlargement of the right heart and its dysfunction which was recognized prenatally, were connected with postpartum mortality¹⁰. Using CVPS to assess cardiac insufficiency could have been valuable information. The prenatal recognition of VGM allows the planning of delivery in a tertiary care health care establishment with an opportunity to urgently transfer the newborn to further endovascular treatment. That is why the proper perinatal care is very important for fetuses with VGM which have signs of cardiac insufficiency.

CONCLUSIONS

1. New techniques of treatment give fetuses with VGM and prenatal signs of cardiac insufficiency a chance of survival and healthy life, in a group of selected cases.

2. The analysis of the cardiovascular system in fetuses with VGM according to uniform criteria is essential in order to assess the influence of these malformations on the survivability of newborns.

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Conflict of interest: The authors declare no conflict of interest

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