Case Report

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# Galen vein aneurysm– challenge for treatment

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**Abstract:** The term vein of Galen aneurysm is related to a group of different vascular anomalies, with one common distinction being the dilatation of the vein of Galen. It represents a rear vascular anomaly, whose incidence is yet unknown, although some authors suggest numbers around 1:25000 deliveries. It accounts for only 1% of all intracranial malformations, but the percentage of this anomaly in pediatric populations is up to 30%. In most cases the diagnosis is made postnatal, while antenatal diagnosis, because of the pathophysiology of aneurysm itself, as well as pathophysiology of its possible complications, is made usually during the third trimester, frequently after the 34th week of gestation. The earliest reported diagnosis was made at 25 weeks' gestation. In this case we present 25-year old gravida, pregnancy at 28th week of gestation with aneurysm, and sonographically detected aneurysm of the vein of Galen, that ended with termination of pregnancy.

**Keywords:** Galen vein; Aneurysm; Vascular malformation

## 1 Introduction

The term vein of Galen aneurysm is related to a group of different vascular anomalies, with one common distinction being the dilatation of the vein of Galen [1]. It represents a rare vascular anomaly, whose incidence is yet unknown [1-3], although some authors suggest numbers around 1:25000 deliveries [4]. It accounts for only 1% of all intracranial malformations, but the percentage of this anomaly in pediatric populations is up to 30% [1,4-7]. In most cases the diagnosis is made postnatal (7), while antenatal diagnosis, because of the pathophysiology of aneurysm itself, as well as pathophysiology of its possible complications, is made usually during the third trimester, frequently after the 34$^{th}$ week of gestation. The earliest reported diagnosis was made at 25 weeks' gestation [2,6,8,9]. This anomaly is considered to be one of the rare causes of cardiac failure [7]. Even though they are rare, these anomalies are of special interest to interventional radiologists, because of potential endovascular treatment which has proven itself to be an effective, and often the only safe therapeutic modality [5], changed generally poor outcome in neonates and infants (high risk of neurological disorders, cardiac failure, high mortality rate up to 90%) and made their chances slightly better nowadays [3,9].

## 2 Case report

25-year old gravida 2, para 1, was referred to our institution. A previous pregnancy was terminated by Caesarian section, with the baby being completely healthy, and current pregnancy was uneventful prior to the last examination. She was referred at 28$^{th}$ week of gestation, by specialist in primary care center, because of suspicion of cardiac anomaly. During her last routine ultrasound check-up, cardiomegaly was noted. Previous check-ups showed regular fetal morphology and biometry.

Biometric parameters (BPD, HC, HL, AC, FL) were corresponding to 25/26$^{th}$ sonar week of gestation, which would correspond to the fetus between 10 and 50 percentile growth.

The 3$^{rd}$ level ultrasound examination revealed a singleton intrauterine pregnancy, in cephalic presentation and placenta located on anterior wall. According to anamnestic data – term of the last menstrual cycle, the pregnancy was at 28$^{th}$ week of gestation (27w 6d), while the estimated fetal weight at the examination as well as other measured parameters (BPD, HC, HL, AC, FL) showed mild fetal growth retardation (24w 5d – 26w 3d), with regular fetal movement and the amount of the amniotic fluid.

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The sonography of fetal brain showed anechoic supratentorial tubular structure, 9,5 mm wide, located sagitally and dorsally above quadrigeminal cistern, spreading bilaterally along the occipital bone, all of which give the appearance of a 'keyhole' (Figure 1). Color Doppler interrogation showed high-velocity vascular turbulence within it, which confirmed the diagnosis of the vein of Galen aneurysm (Figure 2).

Fetal cardiothoracic index was above 0,5 and corresponding to cardiomegaly, with regular 4-chambered cardiac anatomy and normally related great arteries, but with enlarged neck vessels. The dilatation of superior vena cava, ascending aorta and aortic arch branches – brachiocephalic trunk, left common carotid artery and left subclavian artery was noted (Figure 3).

Spectral Doppler showed the increase of velocity through all of the cardiac valves, as well as through the enlarged neck vessels. There was no pleural effusion or tachycardia.

Hepatomegaly without ascites was noted, and there wasn't any change in brain ventricular morphology.

Ten days after, the ultrasound examination was repeated and showed progression in dilatation of large vessels on cardiac base – superior vena cava, the ascending aorta and pulmonic trunk, with dilatation of right cardiac atrium due to increased pressure in dilated superior vena cava. Also, there was the initial dilatation of occipital horns of lateral ventricles, up to 9 mm.

In the case presented here, due to relatively early detection of anomaly – vein of Galen aneurysm (which by itself is a sign of severity), associated cardiomegaly, dilatation of major blood vessels on cardiac base as well as dilatation of neck blood vessels, hepatomegaly and initial dilatation of occipital horns of lateral ventricles, and because of possible postnatal complications, we considered that the prognosis is poor. She was referred to the Committee for prenatal diagnosis in Clinical centre of Montenegro.

Members of the Committee considered that, in case of potential successful completion of pregnancy, chances for healing after embolization therapy were minimal, and since this rare anomaly was presented early in the course of pregnancy in such severe form, which is a sign of intrauterine fetal vulnerability and falls in the category of poor postnatal outcome, termination of pregnancy was the optimal solution and as such was presented to parents. After receiving their approval, the pregnancy was terminated.

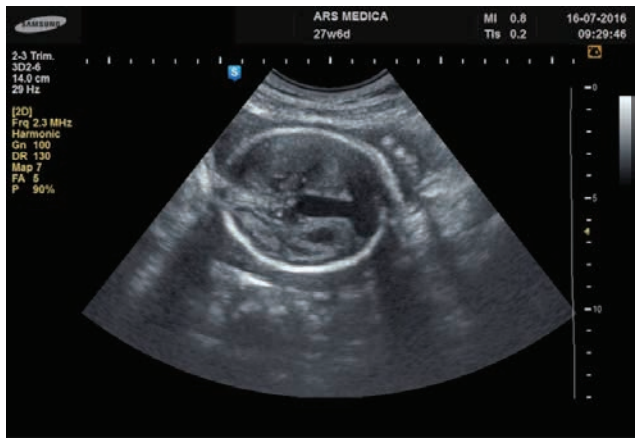


**Figure 1:** Fetal brain transverse view: anechoic supratentorial tubular structure, located sagitally and spreading bilaterally

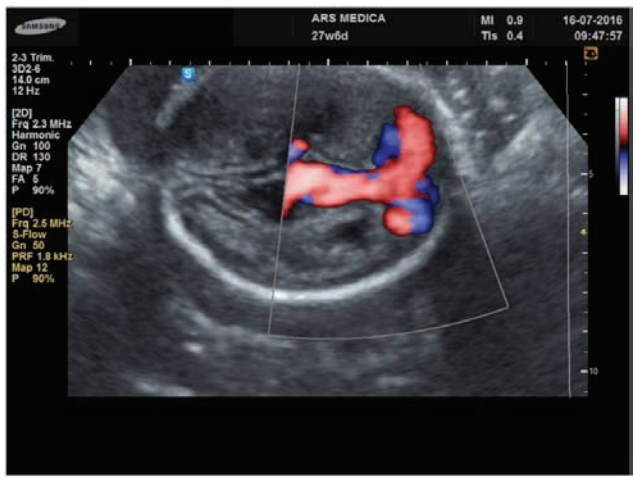


**Figure 2:** Fetal brain transverse view with color Doppler signal: showing turbulent blood flow inside tubular structure

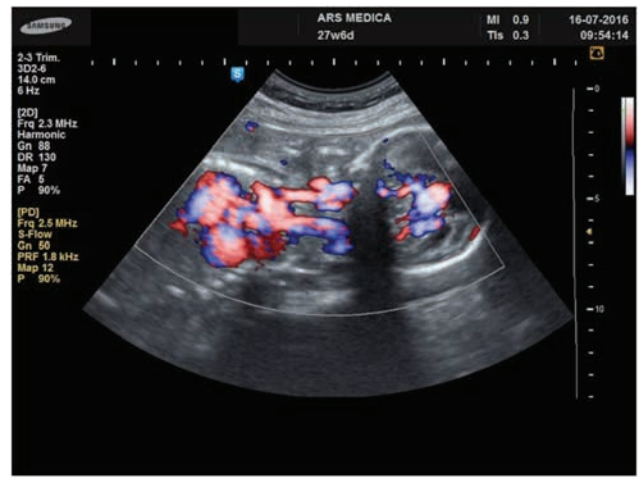


**Figure 3:** Fetal longitudinal parasagittal view: showing cardiomegaly and dilatation of aortic arch branches – brachiocephalic trunk and left common carotid artery

**Consent:** The patient and his family were informed that data obtained would be submitted for publication and a written consent was obtained from the patient and their relatives.

**Ethical approval:** The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

## 3 Disscusion

This malformation was first described in 1895, by Steinheil, referring to it as a 'varix aneurysm'. Since then, it has been variably nominated ('aneurysms of the vein of Galen', 'vein of Galen aneurysmal malformations' 'arteriovenous aneurysms of the vein of Galen'...), but in each described case the common characteristics was the dilatation of the Galen vein in different stages [5,10].

Considering the fact that the vein of Galen aneurysm is a congenital malformation, and also because of better understanding of pathophysiology of the aneurysm itself as well as its complications, the detailed knowledge of its anatomy and embryology is necessary.

The vein of Galen is short (1-2cm long), wide vein, which passes above and behind the splenium of the corpus callosum into quadrigeminal cistern [11]. The vein of Galen is bordered superiorly by the free margin of the falx, posteriorly by the tentorium cerebelli, anteriorly and inferiorly by the roof of the third ventricle, and laterally by the choroidal fissures of the lateral ventricles [1,12]. It is formed by two internal cerebral veins, which again are formed in the level of the interventricular foramen of Monro by union of choroid, septal and thalamostriat vein, and then pass posteriorly along the roof of the third ventricle, 2mm bilaterally from median line, and unite below splenium of the corpus callosum. It receives the basal vein of Rosenthal and the posterior fossa veins and then drains to the anterior end of the straight sinus, at the point where it unites with the inferior sagittal sinus. All venous blood drains into internal jugular veins and then through brachiocephalic veins and superior vena cava into right atrium [11].

The process of cerebral vascularization begins during the 4$^{th}$ week of gestation, at the time of neural tube closure. By the end of the 5$^{th}$ week the main afferent vessels supplying the vein of Galen are formed, and those are choroid and quadrigeminal arteries, while by the end of the 6$^{th}$ week the entire circle of Willis is formed. The anterior cerebral and choroid arteries supply the epithelium of the third ventricle roof, from which the choroid plexus is later formed [1,5,12]. The Internal carotid artery also supplies the diencephalon through posterior communicating arteries, from which the early posterior choroid and mesencephalic arteries are formed. [5,12]. At the same time, while the arteries are being formed, at the roof of the diencephalon the median prosencephalic vein develops as the main drainage vessel of the choroid plexus. By the 10$^{th}$ gestational week it is being replaced, almost entirely, by two internal cerebral veins, except its most caudal part, which then joins the two mentioned veins in forming the vein of Galen [1,5,12]. Sigmoid and transversal sinus are developed by the end of the 7$^{th}$ week [1].

This vascular malformation characterizes the absence of cerebral capillaries, which leads to development of direct communication between cerebral arteries and deep drainage veins, and later on the forming of direct arterial blood shunt into veins, which then consecutively becomes enlarged and aneurismal [13]. Raybaud et al. came to the theory that the beginning of this process happens sometime between 6$^{th}$ and 11$^{th}$ week of gestation, inside velum interpositum and quadrigeminal cistern. Primary `feeders` are considered to be those arteries which normally supply tela choroidea and quadrigeminal lamina, and those include two arterial groups: anterior or prosencephalic (anterior cerebral artery, anterior choroid artery, middle cerebral artery and posterolateral choroid arteries) and posterior or mesencephalic group (posteromedial choroid arteries, posterior thaloperforant arteries, quadrigemina arteries and superior cerebellar arteries), where the main significance have posterior choroid arteries and after them anterior cerebral arteries [12]. After development of AV malformation and occurrence of blood shunt the median prosencephalic vein, as the primary drainage vein, because of lack of fibrous wall and its localization inside the velum interpositum cistern where is largely unsupported, due to sudden high increase in pressure (because of the large amount of blood), becomes aneurismal [5].

The malformation is the most frequently suspected in cases of prenatal ultrasonographic finding of intracranial cystic structure, localized posteriorly, sagitally or slightly parasagitally, supratentorialy, above thalamus (in 92%). When there is simultaneous existence of straight sinus dilatation, the lesion gets a tubular appearance resembling a 'keyhole'. In most cases this malformation is accompanied by sagittal sinus dilatation. The Color Doppler ultrasonography can be used to visualize turbulent blood flow inside

the mentioned cystic formation [2,9]. Differential diagnosis includes arachnoid, porencephalis or choroid plexus cysts, pineal tumors, intraverebral haematoma or choroid papilloma. The diagnosis is confirmed by use of spectral or color Doppler, since none of the mentioned differential diagnosis do show turbulent blood flow, like in cases of the vein of Galen aneurysm. During the ultrasonographic examination of fetuses with this vascular anomaly, one can notice the signs of volume overload, i.e. cardiomegaly, dilated neck venous vessels, as well as hydrops [2,9,14]. Usually to confirm the diagnosis prenatally ultrasound is the method of choice, but for the more precise localization of AV malformation, as well as for identifying which blood vessels are including in its formation, MRI and MRA can be used. The additional diagnostic procedures are usually performed after birth, when precise localization and differentiation of included blood vessels is needed because of embolization treatment.

Sepulveda and al. published the review of known cases of the Vein of Galen aneurysm by that time, in which they write that in 76% of fetuses with this vascular anomaly there were other associated anomalies, and the most common were ventriculomegaly, cardiomegaly (64%), and dilated neck vessels (in 1/3 of fetuses) [9].

The vein of Galen aneurysm is one of the rare causes of cardiomegaly and subsequently cardiac failure in neonates, but if it develops, this cardiovascular complication is one of the leading causes of neonatal and infant mortality and morbidity, where some authors describe neonatal mortality rate over 90% [9]. Cardiac failure can also be a cause of death in utero as well [13,15]. Therefore, even though it is not the most specific signs, especially if it is noticed in third trimester, prenatal cardiomegaly should be a warning sign of possible existence of the vein of Galen aneurysm [9]. In some cases in which the cardiomegaly is present, as much as 80% of total cardiac output is distributed to cerebral blood vessels, because of decreased resistance inside the malformation. This large amount of blood is then distributed back to heart, to the right atrium, which, because of increased cardiac preload, dilates, and then it is followed by dilatations of other cardiac cavities. Diastolic pressure rises, which compromises myocardial perfusion. Because of increased cardiac output, fetal sympathetic tone is augmented, which can lead to sinus tachycardia [1,13]. Ultrasound examination shows dilatation of superior vena cava, right atrium and ventricle, as a result of increased cardiac preload, but also dilatation of aorta and aortic arch branches, with retrograde diastolic flow in arch and proximal part of descending aorta, which can be visualized in color Doppler. Also, Doppler can be used to detect high velocities across all cardiac valves and great vessels [13]. Heart failure usually develops shortly after birth. The most probable explanation for this is relatively high resistance in the cerebral vessels until third trimester together with low placental vascular resistance reduces the amount of blood which is 'stolen' by abnormal low-resistance cerebral shunt [1,3,13]. Of course, the duration of this protective effect depends on the size of AV malformation and subsequent dilatation of the vein of Galen, because in cases of large vascular anomaly, the development of cardiomegaly and heart failure happens earlier, and can lead to hydrops [3].

It can be concluded that the reason for late detection of this anomaly prenatally (usually in the third trimester), or postnatally, probably resides in this possibility of placenta to compensate, up to some degree, the decrease of vascular resistance in AV malformation, as well as the increased blood flow. This can lead to a conclusion that the earlier the anomaly is detected, the worse is the prognosis because the larger size of shunt leads to larger aneurismal dilatation of the vein of Galen, which is then easier to detect earlier in course of pregnancy, but also leads to earlier loss of placenta's defending mechanisms against development of cardiomegaly, heart failure, hydrops and death in utero. Besides, depending on the degree of dilatation, this anomaly can cause the compression of brain tissue, which can lead to slower brain development and cause cerebral atrophy or encephalomalacia, and therefore the earlier detection of this anomaly is also associated with poor prognosis in neurological development of fetus or neonate.

The etiology of hydrocephalus associated with the vein of Galen aneurysm is multifactorial, and includes the Sylvian aqueduct obstruction, resorptive block, hydrocephalus ex vacuo (as a consequence of encephalomalacia and brain atrophy which develop because of a compression of enlarged blood vessel on a developing brain) and abnormal trans-ependymal resorption of cerebrospinal fluid [1,5,7,9,13]. Lately most authors consider that the Sylvian aqueduct obstruction itself doesn't have such an important role in etiology of hydrocephalus, as it is the case an impaired cerebrospinal fluid resorption – resorptive block [5,9]. Considering the fact that arachnoid granulations are still not fully matured, most of the cerebrospinal fluid is being reabsorbed into the cerebral parenchyma so it can be drained via medullary veins, in which there is already higher venous pressure (due to increased blood volume, and restriction in vein drainage), which now results in impaired cerebro-spinal fluid drainage as well and hydrocephalus, but also can lead to cerebral edema and neonatal hypoxia [5].

There are many different classifications of this malformation found in literature. They are made based on multiple criteria: pathophysiology of malformation, localization of AV malformation, the degree of the vein of Galen dilatation (dilatation, varix, aneurismal dilatation...), the combination of previous criteria, as well as based on following complications and possible outcome. We have selected several of them, which are being used most commonly. Yasargil et al classified the vein of Galen aneurysm into four categories based on the pathophysiology of dilatation: type 1 in which there is the direct communication of supplying arteries ('feederes') with the vein of Galen which results in its dilatation; type 2 is composed of thalamoperforators that normally supply the parenchyma, but give branches for communication with the vein of Galen; type 3 is the combination of previous types; and type 4 in which 'feeders' are communicating with one of the proximal veins which drain into the vein of Galen and therefore it is becoming enlarged [16]. Quisling and Mickle describe three types: type 1 is the communication between only one choroidal arterial trunk and the vein of Galen; type 2 is the communication between thalamoperforators and superior thalamic vein which drains into the vein of Galen; and type 3 in which there is a direct communication between multiple anterior and posterior choroid arteries and the vein of Galen [17]. Alvarez and al. recently described type 1 as vein of Galen aneurysmal malformation, known as choroidal type, in which the AV malformation is supplied by the choroidal arteries, and drains via median prosencephalic vein, and it can be diagnosed during the second trimester. Type 2 is vein of Galen aneurysmal dilatation, formed by the AV malformation that drains into one of the usual tributaries of the vein of Galen, and it is usually diagnosed postnatal. Type 3 is vein of Galen varix, in which there is no AV malformation, and type 4 is dural vein of Galen dilatation which is found in adults [18].

The treatment of neonates who present early with complications is still challenging. It is considered that the combination of an early aggressive heart failure treatment and interventional radiological procedure in adequately equipped centers for postoperative care lead to decreased mortality rate. In cases where treatment for failure is unsuccessful, and there is no additional damage in brain parenchyma, early treatment by vascular (arterial or venous) embolization is recommended, otherwise endovascular procedures are delayed until four to six months of age. Prior to that, as mentioned above, MRI with MRA must be performed, because of precise evaluation and localization of AV malformation, as well as to confirm there is no other brain impairment [4,7,19,20].

# 4 Conclusion

The vein of Galen aneurysm is a rear intracranial vascular anomaly, which can, depending on size of AV malformation and consequent shunt, lead to severe complication in utero and early in neonatal period. It is usually discovered in the third trimester, if it is detected earlier there are usually some complications developed as well, and therefore the prognosis is worse. In order to establish the diagnosis of this anomaly, carefully performed ultrasonographic examination of fetal central nervous system is sufficient, especially in the third trimester. Also, if cardiomegaly is discovered, with no other obvious underlying cause, suspicion of this anomaly should exist. The decision for further course of pregnancy, method of delivery, additional diagnostic procedures and therapeutic treatment should be adjusted to each case individually, based on the time of detection of anomaly and presence of associated complication.

**Conflict of interest statement:** The authors certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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