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CASE REPORT

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Prenatal diagnosis of a vein of Galen aneurysmal malformation with MR imaging – report of two cases

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Summary

Background: Vein of Galen malformations (VGMs) are rare congenital defects of cerebral vessels. They are formed between the 6th and 11th week of gestation. The background of this defect involves presence of one or more arterovenous fistulas directing bloodflow toward a persistent, dilated, proximal part of median prosencephalic vein (MProsV). Ultrasound examination is a basic test for diagnosis of VGMs. It has now become possible to acquire images of diagnostic value using magnetic resonance (MR) techniques.

Case Report: This work presents two cases of vein of Galen aneurysms diagnosed prenatally with magnetic resonance imaging. In both patients fetal CNS malformations were diagnosed in ultrasound examinations. MR imaging of the fetal head was performed for further diagnostics.

Conclusions: Because of the ability to precisely determine the size of the ventricular system, presence of raised intraventricular pressure and topographic relationships between pathologically changed vessels and particular cerebral structures as well as the presence of ischemic areas MR examination is currently not only complementary to ultrasonography, but is becoming an independent examination method in the diagnostics of vein of Galen malformations.

Key words: vein of Galen malformations • MRI • median prosencephalic vein

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Background

Vein of Galen malformations (VGMs) are rare congenital defects of cerebral vessels. They constitute up to 30% of intracranial vascular malformations presenting among pediatric patients [1]. They are formed between the 6th and 11th week of gestation. The malformation is due to the presence of one or more arterovenous fistulas directing blood flow toward the dilated, persistent proximal median prosencephalic vein (MProsV). Under normal conditions this vein undergoes regression during embryogenesis before the 11th week of gestation together with development of middle cerebral veins and the vein of Galen [2].

Clinical symptoms depend on the size of arterovenous flow. The most common ones include: antenatal development of heart failure, brain hypoperfusion and hydrocephalus [3].

Ultrasound is a basic examination method allowing for diagnosis of VGMs. Acquiring images of diagnostic value with magnetic resonance (MR) technique has recently become a clinically valuable complementary method [4–6].

In this work we present two cases of vein of Galen aneurysms diagnosed prenatally with magnetic resonance examination performed at the Department of Radiology and Diagnostic Imaging of the Medical University of Lodz.

Case Studies

Case 1

A 22-year-old patient in her first pregnancy. Malformation was diagnosed in an ultrasound examination performed in the 33.4 week of pregnancy outside our Department.

An MRI of the fetal head in T2-weighted sequences in the coronal, sagittal and transverse planes as well as in diffusion sequences was performed in the course of the diagnostic process.

A well-demarcated lesion characterized by loss of signal, 25×21×30 mm in dimensions, was identified near the midline in a projection visualizing the vein of Galen. MRI picture corresponded to a vein of Galen aneurysm (Figure 1A–1C). Loss of signal in the described lesion was strong and homogeneous, indicating a malformation type with direct connection between an artery and a vein. MR image did not allow for determining the vessels that were the source of inflow into the aneurysm and no pathological vessels were identified in immediate proximity of the lesion. However, all vessels of the circle of Willis were dilated, with the basilar artery and posterior cerebral arteries being dilated to greatest extent. Aneurysm was located in the posterior part of the longitudinal fissure, modeling the basal cisterns and pressing against posterior part of the pons. There were no features of brain ischemia noted in a diffusion study. There was no evidence of modeling or midline shift of the ventricular system in the supra- and infratentorial spaces or signs of elevated intraventricular pressure. The ventricular system was of normal size. There were no signs of narrowing the subarachnoid space.

Case 2

A 28-year-old patient in her first pregnancy. A congenital malformation was diagnosed in an ultrasound examination performed in the 33rd week of pregnancy outside our Department.

An MRI examination of fetal head in T2-weighted sequences in the frontal, sagittal and transverse planes as well as diffusion sequences was performed in the course of further diagnostics.

A well-demarcated lesion, 27×22×24 mm in dimensions, located medially and slightly to the left, characterized by loss of signal was identified in a projection visualizing the vein of Galen. MRI picture corresponded to vein of Galen aneurysm. Lesion was directly connected with a dilated (up to 11 mm) straight sinus. Confluence of sinuses including the inferior part of sagittal sinus and both transverse sinuses were also dilated. Basilar artery ran a tortuous course and was noticeably dilated. The remaining arteries forming the circle of Willis were also slightly dilated. The described lesion pressed against the medial part of left hemisphere, slightly modeling the posterior fragment of 3rd ventricle and adhered to the cerebellar tentorium without signs of compression (Figure 2A, 2B). There were no signs of cerebral ischemia noted in a diffusion study. Ventricular system was not dilated, without signs of modeling or compression and without signs of leakage of cerebrospinal fluid. There were no signs of narrowing the subarachnoid space.

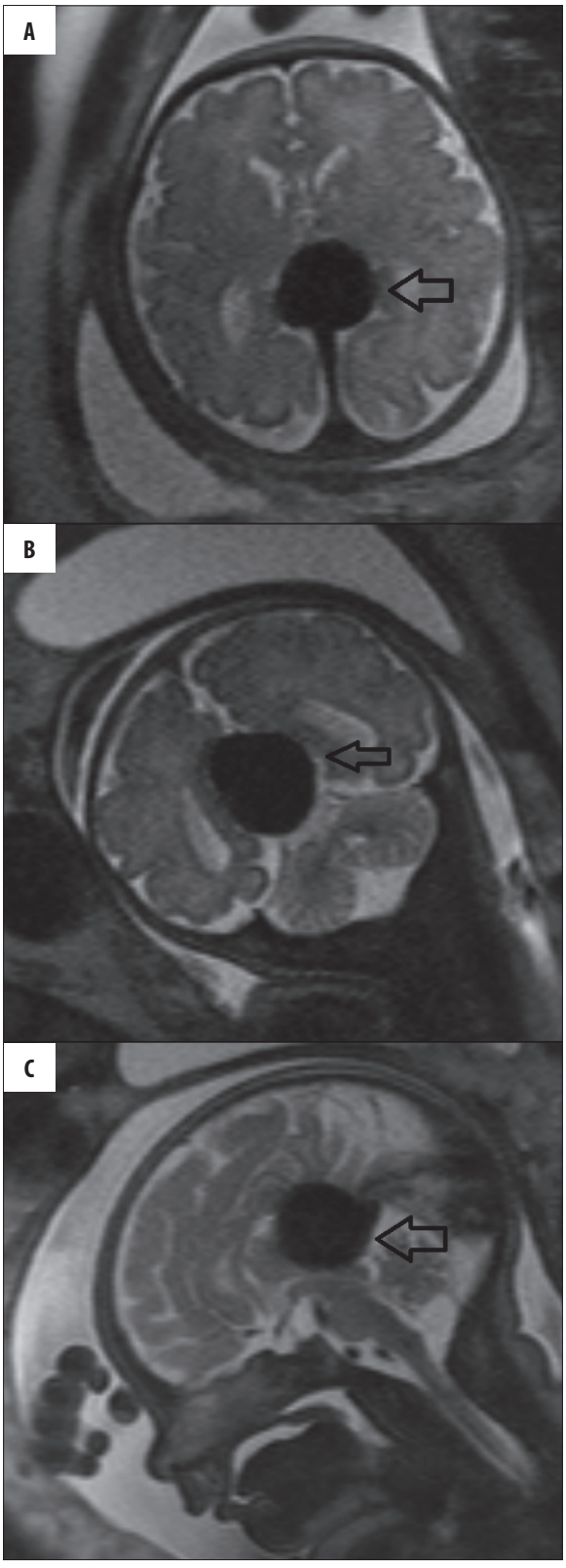

Figure 1. Case 1. Fetal MRI showing the vein of Galen malformation (arrow) (**A**) transverse view, (**B**) coronal view, (**C**) sagittal view.

Discussion

Vein of Galen malformations (VGMs) constitute 1% of all intracranial vascular anomalies. Although rare, they are one of the more frequently arterovenous malformations diagnosed in children both pre- and postnatally [7].

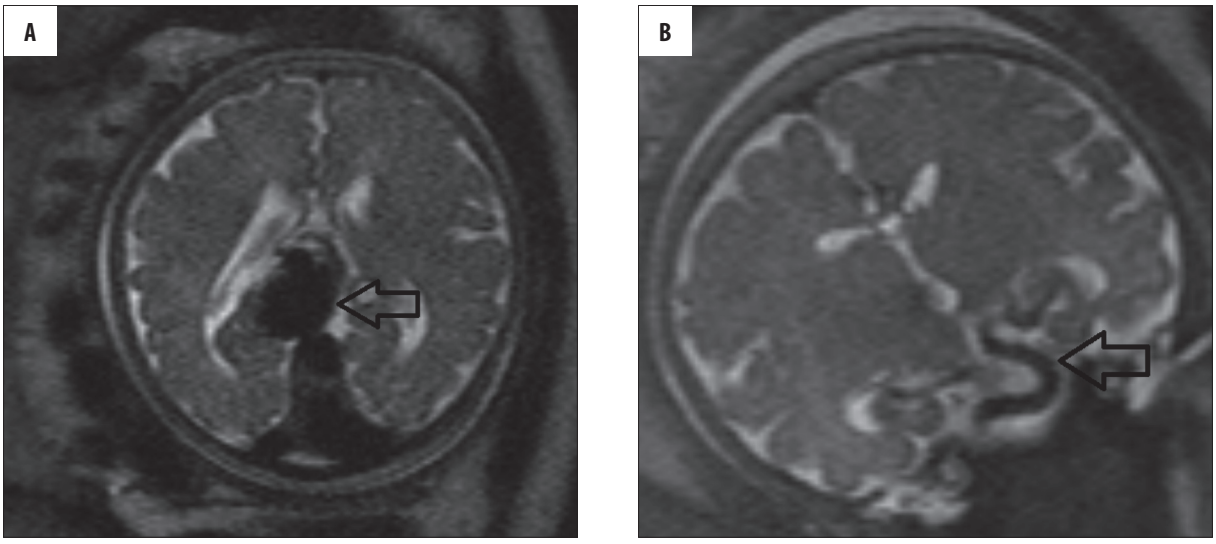

Figure 2. Case 2. (**A**) Fetal MRI showing the vein of Galen malformation on transverse view (arrow), (**B**) coronal view showing dilated and curved basilar artery (arrow).

Table 1. The Yasargil and Lasjaunias classification of vein of Galen Malformation.

	Yasargil classification	Lasjaunias et al. classification
Type I	Fistula between MProsV and pericallosal arteries (anterior or posterior) or PCA	Choroidal type of VGM*
Type II	Numerous fistulas between MProsV and thalamoperforators	
Type III	High-flow type I or II	
Type IV	Arterovenous malformations (AVM) of the mesencephalon involving veins draining into MProsV	Mural type of VGM**

* Blood is supplied to the anterior part of the dilated median prosencephalic vein via venous inflow from fistulas formed between choroidal arteries or other deep mesencephalic arteries. **There is a fistula or fistulas directly in the wall (most frequently lateral) of the median prosencephalic vein. In this type the number of feeding arteries is smaller and the risk of heart failure is lower than in the choroidal type.

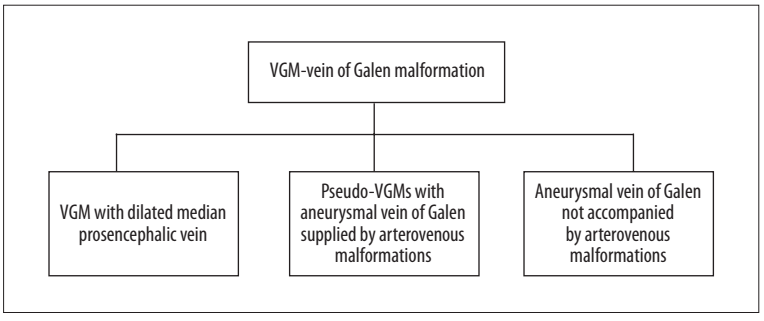

Figure 3. Cerebral vascular malformation classified as vein of Galen malformation (VGM).

There are several other names for this congenital anomaly used in literature such as: "vein of Galen aneurysms", "arterovenous malformations in the vein of Galen", "arterovenous vein of Galen aneurysms", "vein of Galen aneurysmal malformations" [2,7,8]. Nomenclature is quite imprecise, as this anomaly does not involve the great cerebral vein (vein of Galen), but its embryonic precursor – the median prosencephalic vein (MProsV) [7]. In the course of normal development of intracranial vasculature MProsV, also known as Markowski's vein, undergoes involution as middle cerebral veins develop. Its proximal fragment vanishes completely, while the distal part is transformed into the vein of Galen. In the presence of VGMs, formation of arterovenous connections with MProsV not only hinders its regression but lead to significant dilatation of the preserved vessel. In radiological images it presents as a characteristic, dilated structure in the midline of the brain. Moreover, it is also possible that structures typical for early embryonic development such as falcine sinus become preserved, which may result in arresting the development of other sinuses, e.g. straight sinus [9,10].

There are several systems of VGM classification (Table 1). The most commonly used classifications are those by Yasargil and by Lasjaunias et al., categorizing vein of Galen malformations into choroidal and mural based on the

angioarchitecture of cerebral vascular system and location of fistulas [6,11].

Literature refers to three types of anomalies of cerebral vessels as vein of Galen malformations (VGMs) (Figure 3) [12].

VGM drainage and proper venous drainage of the brain are initially separate and intersect at the level of the confluence. Large flow and high flow pressure may lead to formation of venous stenosis. Tolerance of such phenomenon is possible if development is adequate and alternative venous drainage is formed. If venous system is immature, there is increasing compression of brain tissue and arterovenous gradient becomes reduced. It is postulated that presence of VGMs disrupts proper development of intracranial venous drainage [13].

Prognosis in VGMs depends on two main factors. The first one is the severity of heart failure, which is directly related to the size of arterovenous shunt, and the second is the extent of cerebral ischemia caused by increased venous pressure and so-called cerebral steal. Treatment of choice involves performing transarterial embolization in the postnatal period and its efficacy depends largely on the size of malformations and developed complications [14–16].

Ultrasound examination is the study of choice in case of vein of Galen malformations due to its non-invasive character, safety and low cost. Suspicion of VGM is stated during ultrasound examination in the third trimester of pregnancy [4].

Magnetic resonance imaging is a complementary method. It is particularly valuable in cases of ambiguous ultrasound picture in advanced pregnancy, substantial obesity or in oligohydramnios. It is also possible to visualize the fetus in many planes with quite wide field of view [4,17]. What is important, this is a fully objective method. It is also useful in treatment planning in the postnatal period as well as in assessing the prognosis [18,19].

Magnetic resonance technique was first used for visualization of fetal brain in the 80's of the 20th century, although its further development made it possible to fully take advantage of this method in prenatal examinations. However, it is not a screening method due to a relatively low availability, higher cost and lower social acceptability compared to ultrasound examination. There are no reports in the available literature describing negative influence of magnetic field on fetal development. However, small size and usually high mobility of the fetus limit the applicability of MRI in the diagnostics of fetal malformations in the first trimester of pregnancy due to low quality of pictures [20–24].

There is a number of differences between MR examination performed pre- and postnatally. In the first case, we have no control over fetal position and the study itself is often vulnerable to artifacts caused by its movements. Introduction of ultrafast sequences significantly contributed to elimination of this limitation. We also do not perform breath-hold examinations in our facility, as according to our experiences diaphragmatic compression increases fetal movements. Quality of the results depends on proper conduct of the examination and supervision. Before the examination, the patient is placed in a position ensuring the greatest comfort. Duration of the study varies between 20 and 40 minutes [25,26]. An unquestionable advantage of prenatal MR study includes lack of direct exposition of a child to unfavorable conditions of external environment associated with the examination, i.e. noise, low temperature, no need to use sedation) [27].

Currently, MR examinations are most often conducted in a 1.5T field. Shot fast spin-echo (SSFSE) and half-Fourier single-shot turbo spin-echo (HASTE) sequences belonging to T2-weighted sequences are the standard of fetal MR imaging. It produces high-resolution images with small field of view, high contrast-to-noise ratio (CNR) and high signal-to-noise ratio (SNR) [4,26,28].

T1-weighted sequences are also used in prenatal imaging. Most of them are conducted using fast spoiled gradient-recalled (FSPGR) sequences. Longer acquisition times compared to T2-weighted imaging makes these sequences more vulnerable to artifacts ensuing mainly from fetal movements. High water content of the fetal tissues also negatively influences CNR and SNR ratios. T1-weighted sequences are used next to T2-weighted sequences for detection of CNS bleeding, fat-containing structures, i.e. teratoma, and above all assessment of white matter maturity [25,26]. Thus, the role of those sequences in examining vascular malformations is not significant.

Contrary to T1-weighted imaging diffusion sequences (DWI) have a special place in the diagnostics of vascular pathologies. They are based on echoplanar imaging and, due to high usefulness in demonstrating hypoxemic and ischemic changes, constitute an integral part of MRI brain examination [29]. It is particularly important for determination of prognosis and planning of treatment in the postnatal period in case of VGMs. However, one should remember that in case of fetuses below 22nd week of gestation there is a tendency toward acquiring hyperintense DWI signal from the entire fetal body and placenta [30].

MRI studies used in VGM diagnostics do not require the application of contrast agents [31].

Conclusions

In a retrospective analysis, MRI technique was highly valued in the diagnostics of vein of Galen malformations as a study allowing for precise determination of the size of ventricular system, presence of elevated intraventricular pressure, topographic relationships between pathological vessels and particular brain structures as well as the presence of infarcted and ischemic areas [4]. These imaging features are not available in ultrasound examination. Due to its clinical usefulness and safety, MRI study is currently considered a valuable study method in VGM diagnostics [6,20,28].

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