Prenatal Diagnosis of Aneurysms of the Vein of Galen (Vena Magna Cerebri) With Conventional Sonography, Three-dimensional Sonography, and Magnetic Resonance Imaging

Report of 2 Cases

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Objective. To describe the cases of 2 fetuses with aneurysms of the vein of Galen diagnosed prenatally. Methods. The techniques used were conventional sonography, three-dimensional sonography, and ultrafast magnetic resonance imaging. On the basis of these imaging modalities, prognostic factors, such as drainage and secondary damage, were assessed. Results. The first fetus had good prognostic indices, and embolization after birth was successful. According to the prognostic factors, a poor neonatal outcome was predicted for the second fetus. The neonate died soon after birth. Conclusions. Conventional sonography, color Doppler imaging, and magnetic resonance imaging appeared to be useful diagnostic tools. The prognostic factors can be useful in counseling patients and providing the best possible care after birth. Key words: aneurysmal malformations; prenatal magnetic resonance imaging; prognostic indices; sonographic diagnosis; three-dimensional color power angiography; vein of Galen.

Aneurysms of the vein of Galen are rare vascular malformations in children. The actual incidence is unknown but the prevalence is 1% of all neonatal intracranial vascular malformations. The prognosis of infants with vein of Galen malformations in general is rather poor, with a postnatal mortality rate of 50% and a high risk of neurologic disabilities. The arteriovenous malformation should be treated with interventional radiologic procedures, which are preferably performed after the fifth month of life and earlier if the secondary heart failure is untreated.
Prenatally, an aneurysm of the vein of Galen is suspected when a giant midline cystic structure superior to the thalamus is identified on sonography. Different techniques, such as conventional sonography, color Doppler imaging, 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.\(^2,6–9\)

We describe 2 cases of aneurysms of the vein of Galen diagnosed on the basis of the different techniques. This report also shows that prenatal indices can be used to predict the perinatal outcome (Table 1).

**Materials and Methods**

Two patients were referred to our hospital for obstetric sonography. Both patients were sent for evaluation of a cystic structure in the fetal head. The second patient was also referred for evaluation of the heart and growth restriction.

Conventional sonography, color Doppler imaging, MRI, MRA, 3D sonography, and 3D CPA were used to confirm the diagnosis prenatally. In case 1, sonography, MRI, and MRA were also performed after birth and after embolization.

**Table 1. Prognostic Significance of Various Prenatal Sonographic Parameters**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Prognosis</th>
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<tr>
<td></td>
<td>1</td>
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<tr>
<td>Anatomic changes†</td>
<td>+</td>
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<tr>
<td>Hydrops†</td>
<td>+</td>
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<tr>
<td>Dilated drainage tract</td>
<td></td>
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<tr>
<td>Multiple (&gt;5) feeding vessels</td>
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<tr>
<td>Dilated jugular vein/inferior vena cava</td>
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<td>Retrograde aortic flow†</td>
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<tr>
<td>Cardiomegaly</td>
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<tr>
<td>Stenotic or normal-to-minimally dilated straight sinus</td>
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<tr>
<td>No intracranial venous anomaly except vein of Galen malformation</td>
<td></td>
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<tr>
<td>Preserved brain anatomic features</td>
<td></td>
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<tr>
<td>≤2 feeding arteries</td>
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<td>No evidence of high-output state†</td>
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*1 indicates grave prognosis, relative contraindication to intervention; 2, neonatal congestive heart failure, for immediate intervention to prevent intractable congestive heart failure; and 3, benign course, expectant management until the infant is at an age appropriate for embolization, usually 6 months.

†Highly significant prognostic factor.

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**Case Descriptions**

**Case 1**

A 33-year-old healthy gravida 2, para 1 woman was referred to our hospital for fetal evaluation of a large brain cyst detected at 38 weeks’ gestation. The course of the pregnancy had been uneventful before referral. With conventional sonography (2–5 MHz, HDI 5000; Philips Medical Systems, Best, the Netherlands), we depicted a 28 × 19-mm midline tubular anechoic structure situated above the thalamus. Color Doppler imaging identified turbulent blood flow with a velocity of 30 to 40 cm/s and with feeding arteries from the anterior choroidal artery and blood draining into an enlarged straight sinus (Fig. 1). An aneurysm of the vein of Galen was diagnosed. Brain morphologic features appeared normal with no indication of hydrocephaly. The heart was anatomically normal, and there were no signs of hydrops.

With 3D CPA, using the built-in program of the sonography machine (Technos; Esaote-Pie Medical, Maastricht, the Netherlands), the vascular anatomic features were depicted (Fig. 2). The exact course of the dilated supplying vessels and the dilated drainage tract could not be determined. Antenatal ultrafast MRI clearly showed the dilated vein of Galen, which filled the third ventricle almost completely (Fig. 3). At the frontal end, dilated vessels were depicted coming from the choroid plexus on both sides. The straight, transverse, and sigmoid sinuses were enlarged. No structural brain or other abnormalities were seen.

At 39 weeks’ gestation, an elective cesarean delivery was performed. A girl weighing 4860 g with Apgar scores of 9 and 10 at 1 and 5 minutes, respectively, was delivered and transferred to the Pediatric Neurology Department. At physical examination, no abnormalities were found, other than a grade 3/6 systolic cardiac murmur. Echocardiography showed normal left and right ventricle function with mild tricuspid regurgitation. The ductus arteriosus and the foramen ovale were patent with a bidirectional shunt. Brain imaging studies confirmed the prenatal findings. On MRA, the choroidal arteries were depicted as supplying vessels (Fig. 4). Three-dimensional time-of-flight MRA showed arteriovenous malformation of the vein of Galen and enlargement of the sinus rectus without parenchymal abnormalities or hydrocephalus.
On the sixth day, congestive heart failure developed, which responded to fluid restriction and drug therapy. Five days later, the neonate was transferred to the Neonatal Intensive Care unit because of respiratory failure due to congestive heart failure, and mechanical ventilation was necessary. Echocardiography showed severe pulmonary hypertension due to increased pulmonary blood flow. In addition to diuretics, digoxin, and therapy aimed at cardiac afterload reduction, prostaglandin E1 was administered to reopen the ductus arteriosus to reduce pulmonary blood flow.

On the 15th day, the neonate was transferred to a specialized center (Hôpital de Bicêtre, Paris, France) for angiographic embolization of the aneurysm. Embolization of the 2 arterial feeders (=50%) of the arteriovenous malformation was performed on the 19th day. After embolization, she improved considerably. Cardiac evaluation showed normal right and left ventricle function and no signs of pulmonary hypertension. Neurologic evaluation by MRI revealed a substantial decrease of the aneurysmal dilation of the vein of Galen and a small hemorrhage at the trigonum on the right side. Cranial sonography showed mild hydrocephalus, which stabilized spontaneously. The infant's psychomotor development at the age of 3 months was slightly delayed. At the age of 5 months, a second embolization was performed electively. Follow-up at 12 months showed mildly delayed developmental milestones.

Case 2
A 23-year-old gravida 2, para 0 woman was referred for evaluation of fetal cardiomegaly and ventriculomegaly at 33 weeks’ gestation. In her first pregnancy, a bichorial-biamniotic twin pregnancy, unilateral hydrothorax was found in 1 fetus at 14 weeks’ gestation. The hydrothorax dissolved spontaneously at 16 weeks’ gestation. No other malformations were found, and the fetus had a normal karyotype. At 23 weeks’ gestation, both fetuses were delivered premature and died.

The course of the second pregnancy had been uneventful before referral. Advanced sonographic examination of the fetal brain revealed an anechoic tubular structure of 16 × 18 mm located superior to the thalamus. Color Doppler investigation showed bidirectional vascular flow within the structure with velocity of 40-50 cm/s and with feeding arteries from the posterior cerebral artery, and an aneurysm of the vein of Galen was diagnosed. The straight and transverse sinuses were enlarged. The lateral ventricles appeared enlarged with no other structural brain abnormalities (Fig. 5). The heart was mildly enlarged with a cardiothoracic index of 68% without signs of hydrops or pericardial or pleural effusion. Mitral and tricuspid regurgitations were not...
observed. Dilated neck vessels were detected. Fetal biometric measurements were at the fifth percentile for gestational age. With 3D CPA, the supplying vessels were defined as choroidal arteries. It was not possible to determine the course of the drainage tract.

Antenatal MRI (half-Fourier single-shot turbo spin echo) showed a dilated vein of Galen with choroidal arteries as supplying vessels and dilatation of the sinus rectus and sinus confluens. Ventricleomegaly of the lateral and third ventricles was seen, with the vein of Galen filling the third ventricle (Fig. 6). The jugular veins were dilated. Also, the cortical vessels were dilated because of reflux. The parieto-occipital cortex showed possible signs of atrophy.

Because of the cardiomegaly and ventriculomegaly, the prognosis was thought to be poor. Interdisciplinary consultation with the neonatologists, pediatric neurologists, and neuroradiologists confirmed that the chances of successful treatment by embolization of the aneurysm were very low. The parents were informed and decided on conservative management. At 35 weeks' gestation, a girl weighing 1648 g was delivered spontaneously with Apgar scores of 3 and 3 at 1 and 5 minutes, respectively. Immediately after delivery, respiratory failure due to congestive heart failure developed, and the neonate died 40 minutes later. The parents declined an autopsy.

**Discussion**

An aneurysm of the vein of Galen is a vascular malformation of the choroid plexus within the roof of the third ventricle. The dilated venous sac of the aneurysm probably represents persistence of the embryonic median prosencephalic vein. The arteriovenous fistulas, from the choroid arteries, anterior cerebral arteries, transmesencephalic arteries, and other arteries to the vein of Galen, lead to the aneurysmal dilation of the vein. Because of the increased flow and turbulence, the wall of the vein is hypertrophied.

The venous drainage of the vein of Galen normally occurs through the sagittal sinus, which then drains into the confluence of sinuses (the junction of the straight, superior sagittal, occipital, and bilateral transverse sinuses). In cases of aneurysms of the vein of Galen, however, the

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**Figure 3.** Sagittal transverse relaxation time–weighted MRI scan showing a dilated vein of Galen almost completely filling the third ventricle. The straight and occipital sinuses are enlarged.

**Figure 4.** Lateral projection from 3D time-of-flight MRA showing enlarged choroidal arteries feeding the vein of Galen malformation.
straight sinus may be absent, thrombosed, or patent. Then the vein usually drains into the fal
cine sinus, which extends directly into the supe-
rior sagittal sinus.9

Because the blood is shunted to the aneurysm,
hypoperfusion of the cerebral areas, the "steal"
phenomenon, occurs. This hypoperfusion may
result in irreversible ischemic brain damage.1,8

Hydrocephalus is caused by venous hyperten-
sion and alterations in cerebrospinal fluid resorp-
tion or obstruction of the ventricular system.7,8

The placenta protects the fetus by competing
with the aneurysm because it also has low resis-
tance. The amount of blood shunted to the
aneurysm is reduced, and congestive heart fail-
ure is prevented. If the shunt to the aneurysm is
too large, the placenta cannot compete with the
aneurysm, and cardiomegaly and hydrops fetal-
is with brain damage may develop.1,9

Possibilities for diagnostic procedures in
infants with vein of Galen malformations are rel-
atively new. The prognosis for an aneurysm of
the vein of Galen depends on several factors.
Structural brain damage, a dilated drainage tract,
multiple (≥5) feeding vessels, hydrops, a dilated
jugular vein, a dilated inferior vena cava, retro-
grade aortic flow, and cardiomegaly correlate
with an adverse perinatal outcome (Table 1). The
size of the aneurysm is not correlated with the
prognosis. Because the diagnosis is rare, these
indices are based on case reports.1

The first patient had no signs of the steal phe-
nomenon, only a dilated drainage tract. The fetal
growth was normal, and there were no signs of
congestive heart failure. According to the prog-
nostic indices, this fetus had a good prognosis,
and embolization after birth was successful.

The second patient had signs of a dilated
drainage tract, dilated jugular veins, and car-
diomegaly. This fetus was growth restricted, and
fetal movements were reduced, probably
because the largest amount of blood was shunt-
ing to the aneurysm. The brain parenchyma
showed possible signs of atrophy, and ventricu-
locerebral aneurysms slice by slice, but those techniques
did not reveal any additional information. The
fact that 3D CPA did not reveal additional inform-
ation was possibly due to lack of experience
with the technique. The MRI was useful in
excluding cerebral damage and defining the sup-
plying arteries and drainage tract.

Figure 5. Color Doppler image showing bidirectional flow within an enlarged vein
of Galen. The lateral ventricles are enlarged.

Figure 6. Axial transverse relaxation time–weighted MRI scan showing a dilated
vein of Galen, straight sinus, and sinus confluence. The lateral and third ventricles
are dilated. Also note also engorgement of superficial veins.
In conclusion, aneurysms of the vein of Galen and the possible complications can be diagnosed by conventional sonography and MRI. The previously published prognostic indices can be useful in counseling patients and providing the best possible care after delivery.

References


