Prenatal Diagnosis of Vein of Galen Aneurysmal Malformation Allows Early Transumbilical Endovascular Treatment

Diagnóstico pré-natal de malformação aneurismática da veia de Galeno possibilita tratamento endovascular precoce por acesso transumbilical

Zeferino Demartini Jr1,2 Gelson Luis Koppe2,3 Luana A.M. Gatto3,4 Ana Lucia Sarquis4
Alexandre N. Francisco3,4 Adriane Cardoso-Demartini4

1 Department of Neurosurgery, Hospital de Clínicas da Universidade Federal do Paraná (UFPR), Curitiba, PR, Brazil
2 Department of Neurosurgery, Hospital Pequeno Príncipe, Curitiba, PR, Brazil
3 Department of Neurosurgery, Hospital Universitário Cajuru, Pontifícia Universidade Católica do Paraná (PUCPR), Curitiba, PR, Brazil
4 Department of Pediatrics, Hospital de Clínicas da Universidade Federal do Paraná (UFPR), Curitiba, PR, Brazil

Address for correspondence Zeferino Demartini Jr. MD, Complexo Hospital de Clínicas – UFPR, Departamento de Neurocirurgia, Rua General Carneiro 181, 8° andar, Curitiba, PR, Brazil 80060-900 (e-mail: demartiniz@gmail.com).

Abstract

Neonates with vein of Galen aneurysmal malformation (VGAM) presenting with severe cardiac failure and pulmonary hypertension represent a challenge for endovascular therapy. When early treatment is required, the small femoral arteries in this population are usually difficult to cannulate. Alternatively, the umbilical vessels offer a natural pathway to reach the lesion. Therefore, prenatal diagnosis of VGAM allows for delivery planning, perinatal management, and embolization through umbilical approach, thus leading to better outcomes.

Resumo

Neonatos com malformação aneurismática da veia de Galeno (MAVG) apresentando insuficiência cardíaca severa e hipertensão pulmonar representam um desafio para a terapêutica endovascular. Quando o tratamento precoce é necessário, o pequeno diâmetro dos vasos femorais nessa faixa etária dificulta a punção e canulação. Como alternativa, os vasos umbilicais oferecem um acesso natural para alcançar a lesão. Assim, o diagnóstico pré-natal da MAVG proporciona planejamento adequado do parto em local com a estrutura necessária e cateterização dos vasos umbilicais ao nascer, o que permite tratamento precoce e melhor evolução desses pacientes.
Introduction

Neonates with vein of Galen aneurysmal malformation (VGAM) presenting with early congestive heart failure (CHF) have a relatively poor prognosis. The femoral arteries in this population are small and difficult to cannulate; thus, the umbilical vessels are a natural pathway to approach the brain vessels. Nevertheless, the umbilical artery catheterization is best performed right after delivery; therefore, prenatal diagnosis becomes important to plan early treatment and improve outcomes.

Methods and Results

A 29-year-old healthy woman at 31 weeks of gestation underwent a routine fetal ultrasound scan. It identified a 24 × 15-mm midline, anechoic structure above the thalamus. Color doppler revealed a VGAM draining into an enlarged falcine sinus. Fetal echocardiography showed mild cardiomegaly, with enlarged right chambers due to high output. A prenatal magnetic resonance imaging (MRI) during apnea demonstrated a choroidal type of VGAM, with dilated lateral ventricles (Fig. 1). The findings were discussed with the parents, a neonatologist, a pediatric cardiologist, and a neuroradiologist.

At 40 weeks of gestation, a planned Caesarean section was performed, after previous reservation of the hemodynamic suite. The newborn was a 3,250-g male, with normal head circumference; on physical examination, there were a 2/6 systolic murmur and a 3/6 cranial bruit. The Bicêtre neonatal evaluation (BNE) score was 11, while the Apgar scores were 1 at 1 minute, 5 on the 5th minute, and 8 at 10 minutes, after intubation and ventilatory support with oxygen. The neonate developed CHF soon after delivery, which was treated with fentanyl, furosemide, and milrinone. The umbilical artery and vein were cannulated, and he was called for intervention with 23 hours of life. An Echelon 10 microcatheter (Medtronic, Irvine, CA, USA) over a SilverSpeed 10 guidewire (Medtronic) was directly inserted through the umbilical artery cannula, and a digital subtraction angiography (DSA) showed a large VGAM, with several afferences and high-flow arteriovenous fistulas (Fig. 2). Embolization was performed using Axium 3D or Helix mechanical detachable coils (Medtronic) followed by injection of Onyx ethylene-vinyl-alcohol-copolymer (EVOH) (Medtronic) until complete obliteration; the three main afferences were completely occluded, with two being posterior choroidal arteries and one pericallosal. The procedure was interrupted due to contrast volume but reduction of bruit was achieved. In the postoperative period, the infant evolved with tachycardia, treated with esmolol, and pneumonia, treated with oxacillin and amikacin. The cardiopulmonary function progressively worsened, failed to respond to medical treatment using digoxin and sildenafil, and the patient died of intracerebral CHF on the 20th day of life.

Discussion

The VGAM occurs during the 6th to the 11th weeks of gestation, due to the persistence of the median prosencephalic vein (of Markowski). That is the precursor of the cerebral magna vein, which remains connected to the choroidal vessels. Disease expression may vary from several fistulas, inhibiting cardiac function, to complete asymptomatic patients, incidentally diagnosed at adult age.

Prenatal diagnosis of VGAM is commonly made during the third trimester, through fetal ultrasound. Doppler studies can further help to understand the hemodynamics of the lesion, while echocardiography is useful to identify cardiac abnormalities. Fetal MRI in apnea shows large flow void in...
the central region, enlarged falci sine sinus and drainage to transverse sinuses. It also helps to identify hydrocephalus, as shown in our case. The diagnosis during the gestation allows planning of delivery in tertiary hospitals, with high-risk neonatal unit, neurosurgery, and neuroradiology support, favoring early intervention whenever necessary.

Treatment may avoid death and complications, such as hydrocephalus, CHF, or dural arteriovenous fistula. In a meta-analysis of 754 patients with VGAM, 76.7% of untreated patients died; and while microsurgery found an 84.6% mortality rate, endovascular therapy achieved favorable outcome in 72% of the patients, with a mortality rate of 15%. Therefore, embolization is the treatment of choice. It should be attempted as soon as possible for neonates with VGAM presenting CHF and pulmonary hypertension, and a BNE score between 8 and 12 out of 21 requires urgent therapy (Table 1).

The transfemoral approach is commonly used for transarterial embolization; however, catheterization of the femoral artery can be difficult in neonates due to small vessels diameter. Furthermore, it may cause thromboembolic complications or arterial occlusion, and maintenance of a vascular sheath several days for repeated interventional procedures is associated to leg ischemia. The umbilical cord has three vessels: one larger oval umbilical vein, with thin wall, running to the left portal vein; and two smaller, round umbilical arteries, with thick wall, originated from the internal iliac arteries and enabling direct access to aorta. The umbilical arteries suffer prompt constriction after delivery; thus, its cannulation should be conducted immediately after birth; and although it can be performed later, it becomes almost impossible after the 4th postnatal day. Transvenous embolization can be done through direct sinus puncture, jugular, femoral, or umbilical approaches. Because the abrupt total occlusion of the venous side with coils was associated with hemorrhagic complication, we attempted to occlude the high-flow arteriovenous shunts in a graded fashion.

Arterial afferences can be obliterated using n-butyl cyanoacrylate (NBCA), EVOH, coils, or a combination of these. Complications related to embolization of the VGAM include

### Table 1 The Bicêtre neonatal evaluation score (0–21 points) indicates the management

<table>
<thead>
<tr>
<th>Points</th>
<th>Cardiac function</th>
<th>Cerebral function</th>
<th>Respiratory function</th>
<th>Hepatic function</th>
<th>Renal function</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>4</td>
<td>Overload, no medical treatment</td>
<td>Subclinical, isolated EEG abnormalities</td>
<td>Tachypnea, finishes bottle</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>3</td>
<td>Failure; stable with medical treatment</td>
<td>Nonconvulsive intermittent neurological signs</td>
<td>Tachypnea, does not finish bottle</td>
<td>No hepatomegaly, normal hepatic function</td>
<td>Normal</td>
</tr>
<tr>
<td>2</td>
<td>Failure; not stable with medical treatment</td>
<td>Isolated convulsion</td>
<td>Assisted ventilation, normal saturation FiO₂ &lt; 25%</td>
<td>Hepatomegaly, normal hepatic function</td>
<td>Transient anuria</td>
</tr>
<tr>
<td>1</td>
<td>Ventilation necessary</td>
<td>Seizures</td>
<td>Assisted ventilation, normal saturation FiO₂ &gt; 25%</td>
<td>Moderate or transient hepatic insufficiency</td>
<td>Unstable diuresis with treatment</td>
</tr>
<tr>
<td>0</td>
<td>Resistant to medical therapy</td>
<td>Permanent neurological signs</td>
<td>Assisted ventilation, desaturation</td>
<td>Abnormal coagulation, elevated enzymes</td>
<td>Anuria</td>
</tr>
</tbody>
</table>

Abbreviations: EEG, electroencephalogram; FiO₂, fraction of inspired oxygen.

Patient presenting with < 8 points: not to treat; 8–12 points: emergency endovascular intervention; > 12 points: medical management until the child is at least 5 months of age.
neurological disability, death, hemorrhage, and sinus thrombosis.\textsuperscript{1,3} Despite the unfavorable outcome of the described case, in a retrospective review, major brain lesions during prenatal evaluation were associated with poor outcome in all cases.\textsuperscript{2}

**Conclusion**

Prenatal diagnosis of VGAM is important to allow for delivery planning and transumbilical cannulation. This offers a chance for early treatment and improved outcomes.

**Conflict of Interests**

The authors declare that there are no conflict of interests.

**References**


\textsuperscript{4} Marques RM, Lobão CA, Sassaki VS, Aguiar LR. Vein of Galen aneurysm in an adult: case report. Arq Neuropsiquiatr 2006;64(3B):862–864


