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

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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.

Keywords

- vein of Galen malformations
- arteriovenous malformations
- prenatal diagnosis
- umbilical arteries
- newborn infant
- endovascular procedures.

Resumo

Neonatos com malformação aneurismática da veia de Galeno (MAVG) apresentando insuficiência cardíaca severa e hipertensão pulmonar representam um desafio terapêutico. 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.

Palavras-chave

- malformações da veia de Galeno
- malformações-arteriovenosas
- diagnóstico pré-natal
- cordão umbilical
- recém-nascido
- embolização terapêutica

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Introduction

Neonates with vein of Galen aneurysmal malformation (VGAM) presenting with early congestive heart failure (CHF) have a relatively poor prognosis.[1] 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[1] 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 amykacin. The cardiopulmonary function progressively worsened, failed to respond to medical treatment using digoxin and sildenafil, and the patient died of intractable 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).[2,3] That is the precursor of the cerebral magna vein, which remains connected to the choroidal vessels.[2,3] Disease expression may vary from several fistulas, inhibiting cardiac function, to complete asymptomatic patients, incidentally diagnosed at adult age.[4]

Prenatal diagnosis of VGAM is commonly made during the third trimester, through fetal ultrasound.[2] Doppler studies can further help to understand the hemodynamics of the lesion, while echocardiography is useful to identify cardiac abnormalities.[2] Fetal MRI in apnea shows large flow void in

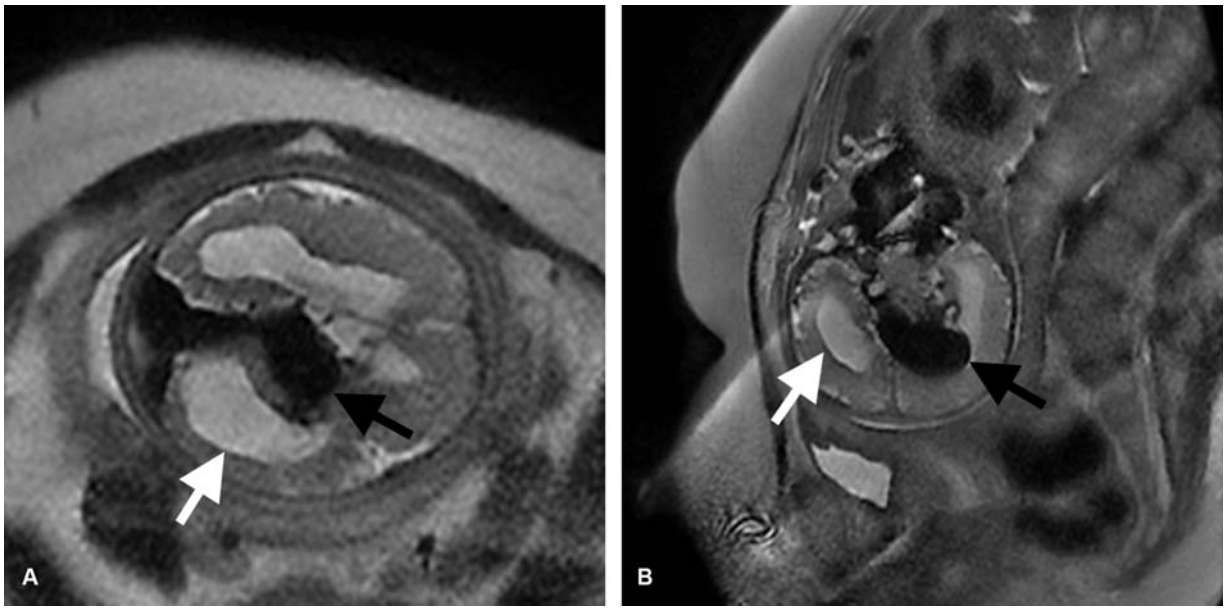

Fig. 1 (**a**) Axial and (**b**) coronal intrauterine T2-weighted magnetic resonance imaging in apnea shows median flow void (black arrows) and lateral ventricles dilatation (white arrows) due to vein of Galen aneurysmal malformation.

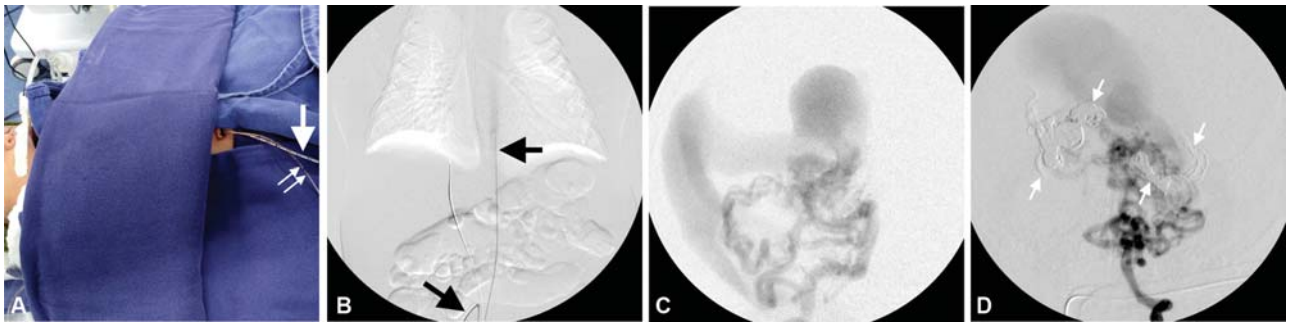

Fig. 2 (**a**) Cannulation of umbilical artery (arrow) and vein (double arrow); the umbilical artery provides easy access to the aorta (arrows) (**b**); vertebral angiogram evidences choroidal type of vein of Galen aneurysmal malformation fed by multiple afferences (**c**); embolization using coils and glue (arrows) occludes the main shunts (**d**).

the central region, enlarged falcine 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%.[5] Therefore, embolization is the treatment of choice.[6] It should be attempted as soon as possible for neonates with VGAM presenting CHF and pulmonary hypertension,[7] and a BNE score between 8 and 12 out of 21 requires urgent therapy (►**Table 1**).[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.[8] 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.[8] Otherwise, the transumbilical arterial approach is technically easier and safer than other methods.[8] 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.[8] 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.[8] Transvenous embolization can be done through direct sinus puncture, jugular, femoral, or umbilical approaches.[8] Because the abrupt total occlusion of the venous side with coils was associated with hemorrhagic complication,[1] 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.[6] Complications related to embolization of the VGAM include

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

Points	Cardiac function	Cerebral function	Respiratory function	Hepatic function	Renal function
5	Normal	Normal	Normal	–	–
4	Overload, no medical treatment	Subclinical, isolated EEG abnormalities	Tachypnea, finishes bottle	–	–
3	Failure; stable with medical treatment	Nonconvulsive intermittent neurological signs	Tachypnea, does not finish bottle	No hepatomegaly, normal hepatic function	Normal
2	Failure; not stable with medical treatment	Isolated convulsion	Assisted ventilation, normal saturation $FiO_2 < 25\%$	Hepatomegaly, normal hepatic function	Transient anuria
1	Ventilation necessary	Seizures	Assisted ventilation, normal saturation $FiO_2 > 25\%$	Moderate or transient hepatic insufficiency	Unstable diuresis with treatment
0	Resistant to medical therapy	Permanent neurological signs	Assisted ventilation, desaturation	Abnormal coagulation, elevated enzymes	Anuria

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.[1]

neurological disability, death, hemorrhage, and sinus thrombosis.[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.[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.

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