# Prenatal diagnosis of intracranial pial arteriovenous fistula
# Diagnóstico pré-natal de fístula arteriovenosa pial

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## Abstract

The vast majority of cerebral vascular malformations reported in the antenatal literature are aneurysmal malformations of the vein of Galen. Other types of intracranial vascular malformations are exceptional in fetuses. The authors present a case of prenatal diagnosis of intracranial pial arteriovenous fistula, not involving the vein of Galen and not showing abnormal intracranial anatomic features, or other sonographic findings.

**Keywords:** Intracranial pial arteriovenous fistula; Non-galenic; Prenatal diagnosis.

## INTRODUCTION

Non-galenic arteriovenous fistula (NGAVF), also known as intracranial pial arteriovenous fistula (AVF), is a rare vascular malformation[1]. Sometimes considered a subset of arteriovenous malformations (AVMs), NGAVFs are distinct in that one or more pial arteries feeds directly into a cortical vein without the intervening nidus of a typical AVM. An NGAVF is distinguished from a dural arteriovenous fistula (DAVF) by the location of the fistula site in the subpial meningeal space and from a vein of Galen malformation (VOGM) by the lack of direct involvement of the embryonic median prosencephalic vein[1]. NGAVF may be located in the posterior fossa or, more commonly, in the supratentorial space. Supratentorial AVFs are always superficial and fed by pial (cortical) arteries[2].

The incidence and prevalence of NGAVF are unknown[1]. In previously published series of pediatric and adult subjects, NGAVF comprised 1.6% and 4.7% of AVM, respectively[3,4]. In two more recent series, NGAVF comprised 7.3% and 8.4% of pediatric brain AVM[1,5]. Since 1970, a total of 168 cases in children and adults have been reported[1]. Assuming that the population prevalence of brain AVM is roughly 10/100,000, then the prevalence of NGAVF can be estimated to be 0.1-1/100,000[1].

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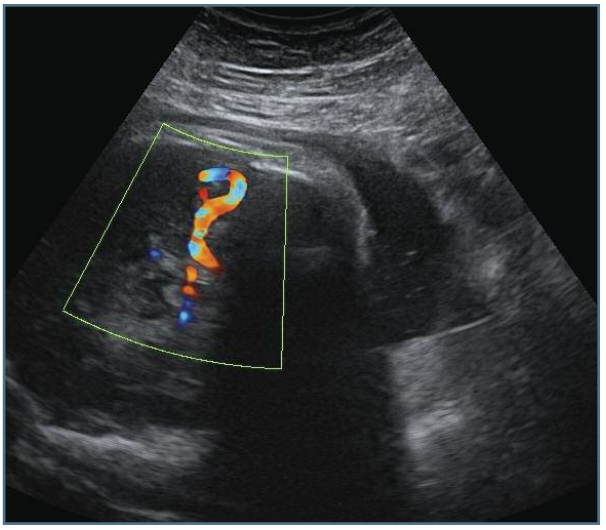
Cerebral arteriovenous malformations are rarely diagnosed in utero. Most prenatal imaging of intracranial vascular malformations relates to VOGM or DAVF. As far as we know, NGAVF revealed in the antenatal period, without aneurysmal dilatation of the vein of Galen, signs of cardiac decompensation or other sonographic findings, has been reported only once and appeared as an anechoic lesion on the prenatal ultrasound scan[6]. The authors hereby present, for the first time, a case-report of prenatal diagnosis of NGAVF, not involving the vein of Galen and not showing abnormal intracranial anatomic features or other sonographic findings on two-dimensional ultrasonography, using color Doppler ultrasonography.

## CASE-REPORT

A 27-year-old woman, primigravida, was referred to our institution for ultrasound examination, at 36 weeks gestation, to estimate fetal weight for suspected macrosomia. The course of pregnancy had been uneventful up to that time. She had Hereditary Hemorrhagic Telangiectasia, previously subjected to several embolization treatments for pulmonary AVMs, with other first degree relatives also affected. Two-dimensional ultrasonography revealed no abnormal intracranial anatomic features, or other sonographic findings, and a harmonious fetal growth above the 90$^{th}$ percentile. Intracranial color Doppler ultrasonography showed an abnormal configuration of the left middle cerebral artery, with increased caliber and turbulent flow, in the upper part of the Syl-



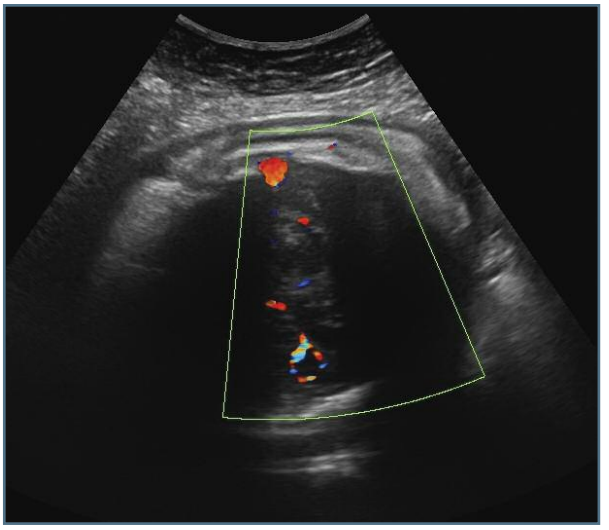


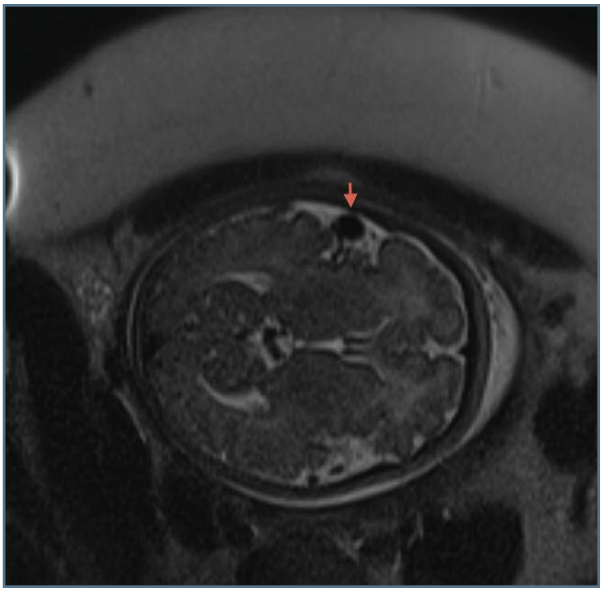
**FIGURE 1 and 2.** Obstetric sonographic examination at 36 weeks gestation, demonstrating an intracranial high-flow pial AVF using color Doppler

vian fissure, and low resistance index, RI 0.54 (Figure 1--2). Taking into account the clinical history and sonographic findings, it was considered the diagnostic hypothesis of an AVM. The final diagnosis of pial AVF was established by fetal magnetic resonance imaging (MRI), performed at 37 weeks (Figure 3-4). There were no signs of cardiac failure or brain damage in subsequent scans and an elective cesarean section was performed at 39 weeks, for suspected fetal-pelvic disproportion and fetal contraindication for instrumental vaginal delivery (fetus with a potentially hemorrhagic condition: AVF grade IV, Cognard system, with 65% hemorrhagic risk[7]). A 4060g female newborn was delivered uneventfully. On the first day of postnatal life, cardiac ultrasound was normal and transfontanellar Doppler ultrasonography confirmed the antenatal diagnosis. The baby was discharged on the 12$^{th}$ day of life, asymptomatic; MRI performed at 3 weeks of age, classified the pial AVF as a single-hole fistula. A multidisciplinary team, involving highly differentiated experts from international centers of reference, prepared a safe and appropriate therapeutic strategy. At six months of age, the child was submitted to embolization of the AVF with success, at Bicêtre Hospital (Paris, France), with no complications and normal development to the date.

## DISCUSSION

Hereditary Hemorrhagic Telangiectasia (HHT; Osler--Weber-Rendu Syndrome) is a relatively common genetic condition affecting 1 in 5–8000 Europeans. While the vast majority of pregnancies of women with HHT proceed uneventfully, the mortality rate is significantly increased due to pulmonary AVM (PAVM) hemorrhage, strokes and myocardial infarction. Thus, all pregnancies with HHT should be considered high risk and women should be advised about the small but serious risks, though reassured that these appear less threatening where medical and obstetric services are aware of the complications. In general, for HHT--affected adults, screening and treatment of PAVMs is recommended to reduce later complications such as paradoxical embolic stroke and hemorrhage, but PAVM screening and treatment of asymptomatic individuals during pregnancy is not justified. For the majority of women with HHT, no cerebral MRI screen is recommended, unless for women with cerebral symptoms, from cerebral hemorrhage. Exclusion of spinal AVMs by MRI scan during pregnancy should be done, to allow regional anesthesia. As for delivery is concerned, prolonged second stage of labour in women in whom cerebral AVMs have not been excluded should be avoided. Hemoptysis of any degree or sudden severe dyspnea should be considered a medical emergency, prompting urgent hospitalization and institution of appropriate treatment[8].

Pial AVFs are rarely diagnosed in utero and this may be due to a lack of understanding of the condition and because AVFs may be peripheral and hard to visualize[2].



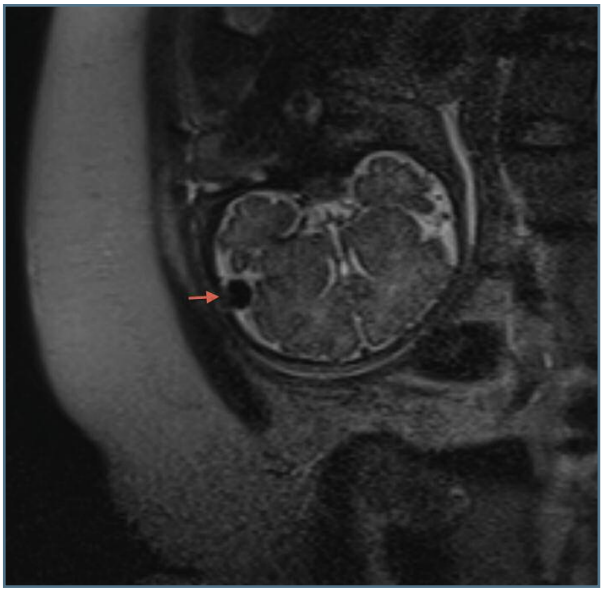


**FIGURE 3 and 4.** Fetal brain MRI, T2-HASTE weighted transverse and coronal images, showing vascular ectasia pericerebral (14x10x12mm), in the upper part of the left Sylvian fissure, accompanied by left middle cerebral artery with increased caliber and dilatation of the adjacent cortical veins (Trolard and Labbé), thus, pial AVF, with the contribution of branches from the left middle cerebral artery. The authors highlight the nonexistence of mass effect on the brain parenchyma, edema or ischemia of venous origin

Intracranial AVM in the fetus reported in the prenatal series and case reports were mainly aneurysms of the vein of Galen. They appeared on the real-time ultrasonography as abnormal intracranial structures such as cystic or tubular structures. Different Doppler means (pulsed, color, power and three-dimensional power Doppler ultrasonography) were then used to confirm the vascular origin of the lesion. Additional associated findings were typically signs of high cardiac output due to the AVF and included dilated neck vessels, cardiomegaly, valve regurgitation and, in some cases, polyhydramnios[8]. The case we report here was an intracranial AVF in a fetus not involving the vein of Galen, not showing abnormal intracranial anatomic features or other additional findings, on two-dimensional ultrasonography. Only screening of the head with color Doppler ultrasonography enabled the detection of the AVF. The routine use of color Doppler ultrasonography in fetal malformations has been discussed controversially and only few reports have supported its clinical utility in the assessment of malformations[9]. Some malformations of the fetal brain are often associated with an abnormal course of intracranial vessels and their visualization using color Doppler can be used for confirming the diagnosis. This report shows that the anomaly was detectable only by color Doppler and not only supported by it, as reported in cases of aneurysms of the vein of Galen, emphasizing the importance of its applicability when suspecting for vascular malformations. We, therefore, consider that any pregnant woman who has a personal or strong familiar history of AVF should have a Doppler fetal brain scan during pregnancy.

AVF is a condition with high morbidity and mortality. Patients who present in the first two years of life are more likely to harbor large, complex multi-hole AVFs that have high flow arteriovenous shunting leading to congestive heart failure (CHF). Children presenting after two years of age were more likely to have a single-hole AVF and to present with seizures, focal neurologic deficits or intracranial hemorrhage. Neonatal NGAVFs are particularly difficult to treat[1]. Embolization is the treatment of choice[1,2,5] and demonstration of the location and nature of the AVF, the arterial supply and number and location of draining veins can be anticipated by angio-MRI prior to the invasive procedure. Parents should be counseled, however, that this is a high-risk procedure for neonates and that fatal complications may occur[1,2,5]. Similar to the management of VOGMs, delaying NGAVFs treatment, until after the first several months of life, is recommended, unless CHF necessitates early intervention[1]. The

goal of eliminating arteriovenous shunting is not for the prevention of CHF, but rather to reduce chronic cerebral venous hypertension that can lead to white matter calcification and "melting brain" syndrome[1], allowing normal neurocognitive development and normal brain maturation[2].

Prenatal detection and precise description of a vascular lesion can, thus, improve postnatal outcome by preparing neonatologists and interventional radiologists to organize an optimized postnatal diagnosis and appropriate therapy[9].

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