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Prenatal diagnosis of intracranial pial arteriovenous fistula and endovascular treatment during the neonatal period

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ABSTRACT
We present a case with prenatal diagnosis of an intracranial high-flow pial arteriovenous fistula that was draining into the vein of Galen in the third trimester of pregnancy. The child was treated by transcatheter embolization with N-butyl 2-cyanoacrylate (NBCA) via the umbilical artery in the early neonatal period due to intractable cardiac failure. Hydrocephalus developed and a ventriculoperitoneal shunt was placed. At the time this report was prepared, the patient was 20 months old and without cardiac failure, but with a delay in neurological development. Prenatal diagnosis and endovascular treatment in the early neonatal period is important in preventing heart failure and resultant mortality due to such high-flow vascular malformations. To the best of our knowledge, the combination of prenatal diagnosis of an intracranial high-flow pial arteriovenous fistula draining into the vein of Galen and endovascular treatment in the early neonatal period is presented here for the first time.

Key words: • *arteriovenous fistula* • *ultrasonography* • *prenatal diagnosis* • *interventional procedures*

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Intracranial pial arteriovenous fistulas are a very rare congenital abnormality that can cause severe morbidity and mortality, particularly in neonates. Congenital arteriovenous fistulas tend to have larger connections and increased flow. The etiology remains unknown. Pediatric arteriovenous fistulas often present in the neonate, suggesting that they are developmental (1, 2). They can present much like a vein of Galen malformation. Neonates usually present with fatal high-output cardiac failure. Medical treatment is inefficient, because of the large, persisting systemic shunt. Mortality is high in the neonatal period if no treatment is offered (1). Emergent diagnosis and management of this pathology is crucial (3, 4). Antenatal diagnosis has been associated with improved outcome (5, 6). Surgery offers little improvement, with fatal outcomes in 80% to 100% of cases. Recently, results have improved markedly with endovascular management (2).

Case report

A 30-year-old female underwent obstetric ultrasound examination in the 34th week of her pregnancy. ultrasonography demonstrated hyperechoic parenchyma of the right parietooccipital lobe, dilation of the vein of Galen, and a straight sinus, all consistent with an intracranial high-flow pial arteriovenous fistula draining into the vein of Galen (Figure 1). Fetal biometry was consistent with the 34th week. Prenatal sonographic findings such as polyhydramnios or hydrops fetalis were not detected.

The mother refused to transfer to a facility with endovascular treatment capabilities before the delivery. She also refused to have a cesarean section, which was proposed in order to minimize cardiac risks to the infant. After uneventful spontaneous vaginal delivery and stabilization of the neonate, contrast-enhanced cranial computed tomography (CT) was performed. Diagnosis of a right parietooccipital lobe arteriovenous fistula draining into the vein of Galen was confirmed (Figure 2). CT also demonstrated the dilatation of the vein of Galen and the straight sinus, right parietooccipital atrophy, and an extended ischemic lesion.

The patient was then referred to a facility with endovascular treatment capabilities due to medically intractable congestive heart failure. After percutaneous umbilical arterial catheterization, a diagnostic cerebral angiography and endovascular treatment procedure were performed under general anesthesia. An arteriovenous fistula supplied by both carotid and posterior cerebral circulation that was draining into the vein of Galen was observed (Figure 3). Multiple feeding vessels originating from the right anterior and posterior cerebral arteries were catheterized with microcatheters and embolized with N-butyl 2-cyanoacrylate (NBCA) (Histoacryl, B/Braun, Melsungen, Germany) diluted with iodized oil (Lipiodol). The NBCA/Lipiodol ratio was 1:1. In total, 2 ml of this mixture was injected through the distal pericallosal branch of the right anterior cerebral artery

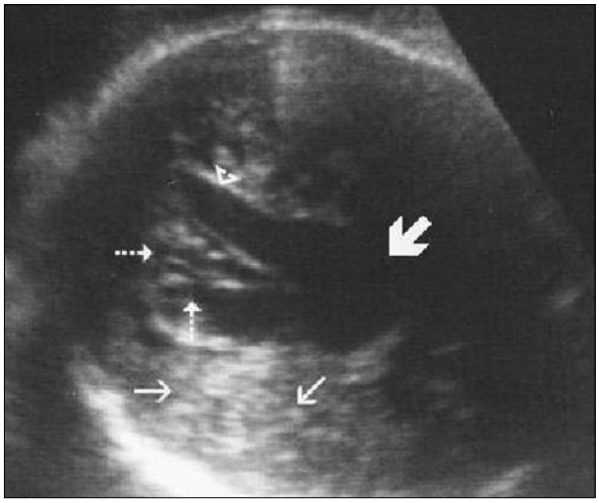

Figure 1. Obstetric sonographic examination demonstrates an intracranial high-flow pial arteriovenous fistula *(dashed arrows)*, draining into the vein of Galen. Hyperechoic parenchyma of the right parietooccipital lobe *(thin arrows)*, dilated vein of Galen *(thick arrow)*, and straight sinus *(arrow head)* can also be seen.

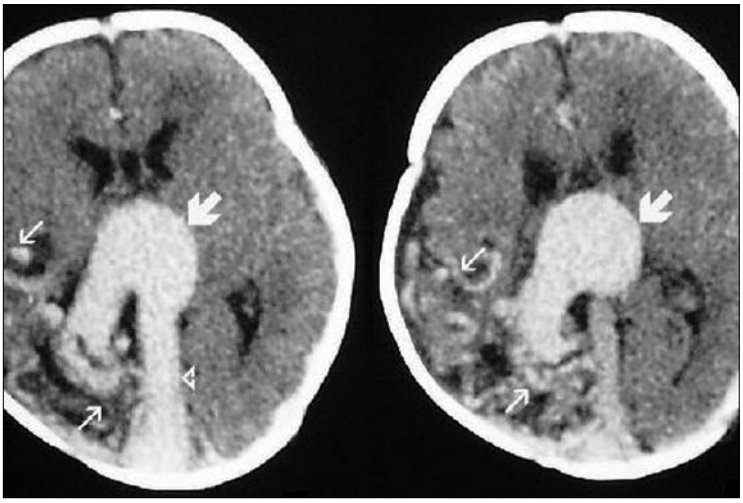

Figure 2. Sequential postnatal contrast-enhanced CT images demonstrate right parietooccipital lobe arteriovenous fistula *(thin arrows)* draining into the vein of Galen, the dilatation of the vein of Galen *(thick arrows)*, the straight sinus *(arrow head)*, right parietooccipital atrophy, and ischemia.

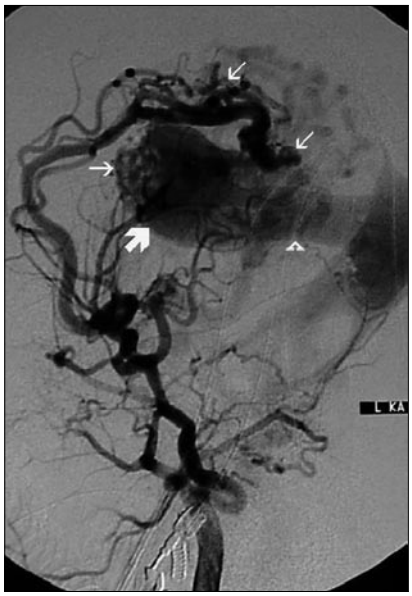

Figure 3. Left carotid artery angiogram demonstrates arteriovenous fistula *(thin arrows)* draining into the vein of Galen *(thick arrow)*. Straight sinus *(arrow head)* can also be seen.

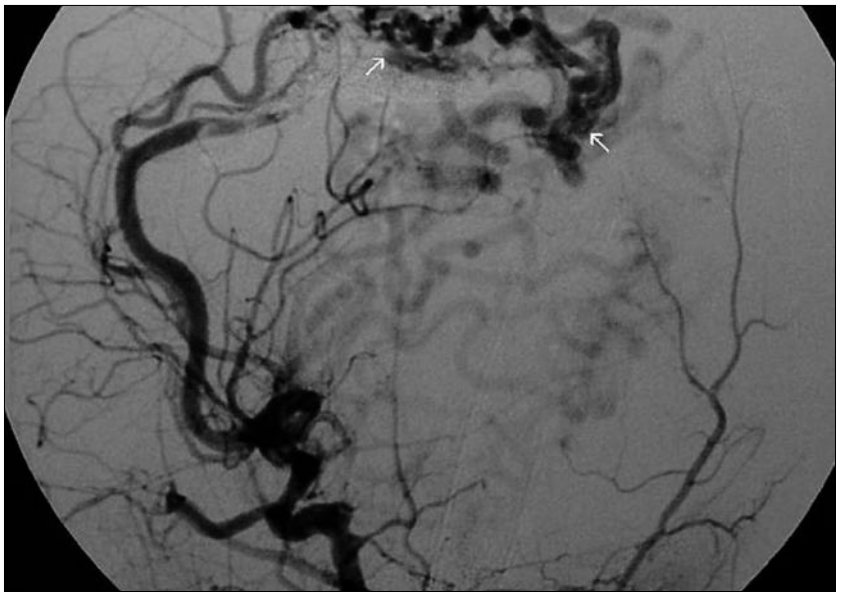

Figure 4. Post embolization angiography demonstrates almost complete angiographic obliteration of the arteriovenous fistula *(arrows)*.

and right posterior choroidal artery, which is a branch of right posterior cerebral artery. Anticoagulation during the procedure was performed with an IV bolus injection of heparin, 100 U/kg. Additional heparin doses were given to maintain activated clotting time (ACT) at around 300 seconds. Heparin was discontinued immediately after the procedure. The patient was 2 days old at the time of embolization. Post-embolization angiograms showed near-complete angiographic obliteration of the arteriovenous fistula (Figure 4). A venous route was not preferred due to the existence of large multiple arterial feeders. Ease of umbilical arterial catheterization was also another factor for preferring an arterial route. Follow-up cranial CT examination, which was performed 1 month after the procedure, showed an occluded arteriovenous fistula and obstructive hydrocephalus. A ventriculoperitoneal shunt was placed due to refractory hydrocephalus. At 20 months old, the patient was without cardiac failure, but with a delay in neurological development.

Discussion

Intracranial pial arteriovenous fistulas are rare vascular lesions, of which fewer than 90 cases have been reported in the literature (7). They are frequently associated with increased blood flow through the midline venous system, causing aneurismal dilatation of the vein of Galen (1, 8, 9). In the reported cases, the prenatal diagnoses of arte-

riovenous fistulas have usually been made during the third trimester (8, 10). The main differential diagnosis is the vein of Galen aneurysmal malformation (VGAM). Lasjaunias et al. have separated VGAMs from the vein of Galen aneurysmal dilatations (VGADs) (11). VGADs have parenchymal arteriovenous malformations that drain through the vein of Galen. Differential diagnosis also includes cystic lesions of the fetal brain, such as arachnoid cysts, choroid plexus cysts, choroid papillomas, porencephalic cysts, pineal tumors, and intracerebral hematomas (8).

The intrauterine diagnosis of this vascular anomaly has been facilitated by the widespread use of ultrasonography and color Doppler sonography, by identifying dilated veins and arteriovenous shunts with turbulent flow (1-5). According to some studies focused on prognostic criteria, the perinatal prognosis seems to be invariably compromised when signs of cardiac decompensation develop prenatally (5, 6).

Cessation of the blood flow at birth to a low resistance territory, such as the placenta, will increase the intracranial vascular shunt, producing hemodynamic decompensation, cardiac failure, and death (8). Damage to the brain may occur, depending on the volume of blood that is shunted through the malformation and away from the brain parenchyma. This redirection of blood flow, known as the steal phenomenon, ultimately causes brain ischemia (12).

Most of the morbidity and mortality related to this anomaly is due to cardiac failure in the neonatal period. The high incidence of cardiomegaly in neonates with arteriovenous malformations also suggests that high-output cardiac failure is already present during the third trimester in a significant number of cases (13). When an arteriovenous malformation is large enough to be detected prenatally, it is likely that it will lead to cardiac failure, either during the antenatal period or just after birth (1). The prognosis in prenatally diagnosed cases has been uniformly poor (8, 14).

This pathology has different clinical presentations. Neonates can present with intractable cardiac failure and intracranial bruit. In infancy, it can present with hydrocephalus and seizures. Older children or young adults can present with intracranial hemorrhage (1).

Due to developing endovascular techniques, the prognosis of neonates with arteriovenous fistulas has markedly improved. Endovascular therapy with modern neuroanesthetic and neurointensive care can provide good outcomes, even in the highest-risk neonates with arteriovenous fistulas and cardiac failure. Medical management alone can rarely control cardiac failure. If medical management of cardiac failure does not succeed, and if there is no evidence of prominent cerebral parenchymal damage on imaging, emergent endovascular treatment is feasible and can reduce mortality (3, 15). Transarterial embolization is the preferred technique, and is more effective in controlling heart failure when there is only 1 or a limited number of arterial pedicles (16, 17). When there are numerous small arterial feeders, it is often impossible to achieve occlusion of more than 1 or 2 of these feeders in the unstable neonate. In this situation, a transvenous embolization route can be used. The transvenous approach can be easily repeated multiple times and may be supplemented by transarterial embolizations. Sonographically guided percutaneous transtorcular embolization is also reported (2). Although the ultimate goal is to obliterate the fistula completely, partial occlusion of the fistula is also very beneficial for the improvement of cardiac function (1, 2, 18, 19).

Umbilical catheterization is associated with some complications, most frequently infection and thrombosis. None of these was observed in our patient due to short-term catheterization (20). We believe the follow-up protocol should involve CT and angiography, in addition to clinical follow-up. CT can show hydrocephalus and parenchymal changes, as it did in our case. Angiography is also necessary to confirm the occlusion or patency of the fistula.

With the development in skills of interventional radiologists, improved embolization technologies and tools, and the noninvasive nature of endovascular procedures, the role for surgery has diminished (1, 2). Nevertheless, if interventional techniques cannot reach or obliterate significant amounts of the feeding vessels, operative clipping may be indicated (21).

The management of neonates with arteriovenous fistulas remains challenging, and is best achieved by a multidisciplinary team that includes an interventional neuroradiologist, neurosurgeon, neonatologist, and anesthetists working in synergy (2).

In conclusion, emergent diagnosis and treatment of this pathology is crucial due to its associated high morbidity and mortality. Endovascular techniques can provide safe and successful treatment for these infants.

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