

Huge cerebral pial arteriovenous fistula in a newborn: illustrative case

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BACKGROUND Pediatric arteriovenous malformations (AVMs) and pial/dural arteriovenous fistulas (AVFs) are rare but life-threatening complications that can lead to congestive heart failure and hemorrhagic stroke in newborns and pediatric patients. The pronounced shunting in these conditions is associated with early complications and necessitates aggressive surgical management. Here, the authors describe endovascular treatment of an atypical cerebral pial AVF in a newborn.

OBSERVATIONS This AVF formed direct communication between a major cerebral artery (basilar artery) and a large draining vein (dilated deep cerebral vein). The authors performed earlier subtotal embolization of the AVF using 0.020-inch coils, which led to progressive thrombosis of the fistula with restoration of normal arterial blood flow. The patient was discharged 18 days after surgery, examination at 1.5 and 6 months showed magnetic resonance imaging signs of blood flow absence through the fistula and satisfactory condition of the infant without physical and mental developmental delay.

LESSONS Subtotal coiling of a high-flow pial AVF in a newborn can result in a good clinical outcome.

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KEYWORDS coil; embolization; newborn; cerebral; arteriovenous; fistula

Pediatric cerebral pial fistulas are a rare cerebrovascular pathology with an incidence of 0.1–1 case per 100,000 population.¹ They belong to a large group of pediatric cerebrovascular pathologies that includes arteriovenous (AV) shunt diseases such as arteriovenous malformation (AVM) of the vein of Galen and dural/pial arteriovenous fistulas (AVFs), venous malformations (including cavernomas), and proliferative diseases (hemangiomas, proliferative angiopathy, and moyamoya disease).² AVMs of the vein of Galen and pial/dural AVFs are the most life-threatening conditions because they can lead to congestive heart failure and hemorrhagic stroke in newborns and pediatric patients. In this case, more pronounced shunting is associated with early complications, whereas the risk of adverse outcomes reaches 90% in the natural course of the disease. This necessitates aggressive surgical management in patients with

this pathology, in particular in newborns.^{3,4} Our experience in the endovascular treatment of cerebrovascular pathology in newborns and children under 3 years of age is more than 60 operations in 35 patients.⁵ However, in the presented case, we report not a typical AVF in which afferent vessels are second- and third-order arteries but an AVF with direct communication between a major cerebral artery (basilar artery) and a large draining vein (dilated deep cerebral vein).

Illustrative Case

History and Examination

The mother was a young woman aged 26 years with an unremarkable obstetric and gynecological history. This was her first pregnancy.

ABBREVIATIONS AV = arteriovenous; AVF = arteriovenous fistula; AVM = arteriovenous malformation; MRI = magnetic resonance imaging; PCoA = posterior communicating artery.

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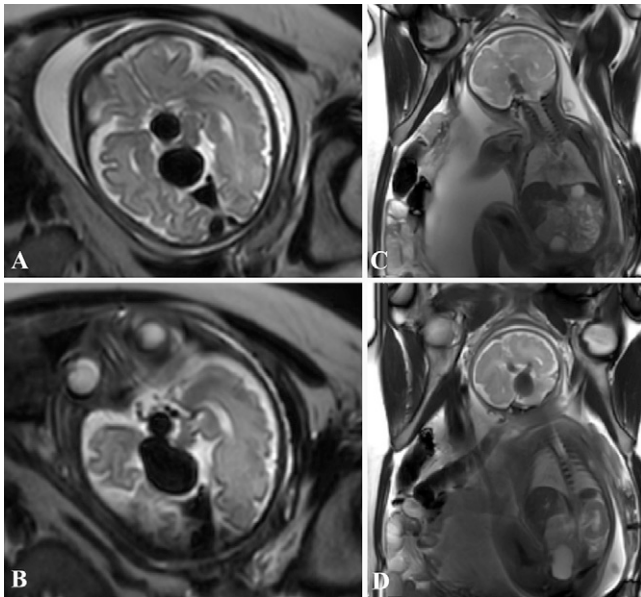


FIG. 1. MRI scans of a 30-week fetus. A dilated venous sac and a fistula between the basilar artery and the deep cerebral vein are seen as black signal voids in axial (**A and B**) and coronal (**C and D**) balanced turbo field echo images. A dilated vein of Galen and a straight sinus are also visible.

The first indirect signs of cerebrovascular pathology of the fetus were detected at the second ultrasound screening (week 20 of pregnancy). The third ultrasound screening (week 29 of pregnancy) confirmed the presence of a large intracranial vascular malformation. Fetal 1.5-T magnetic resonance imaging (MRI) was performed at week 30 of pregnancy on a 3-T scanner (Ingenia, Philips Healthcare) and confirmed a

huge AVF at the skull base with no evidence of encephalomalacia and hydrocephalus (Fig. 1).

After intrauterine examination of the fetus, pregnancy continuation and cesarean section were recommended. Emergency intervention was indicated after birth.

The mother had a routine delivery of a full-term infant at a gestational age of 38 weeks 3 days; a Pfannenstiel incision and a cesarean section in the lower uterine segment with a transverse incision were performed. The delivery occurred without complications; a male with a birth weight of 3,010 g, a birth height of 51 cm, and an Apgar score of 7/8 was born.

The infant was transferred to the neurosurgical department the next morning. The patient's Bicêtre score was 11. Considering the pronounced AV shunting through the fistula, severe pulmonary hypertension, and an unfavorable prognosis of the natural course of the disease, emergency endovascular treatment was indicated for the patient. The newborn was also diagnosed with nonsyndromic congenital heart defects: a 5-mm secondary atrial septal defect, a 2-mm muscular ventricular septal defect, and a 2.2-mm spontaneously closing patent ductus arteriosus. The estimated pulmonary artery pressure was 58 mm Hg, which indicated a significant effect of the fistula shunting to the superior vena cava system and excessive pulmonary blood flow with pulmonary hypertension and the risk of right ventricular failure. Examination by a cardiac surgeon did not reveal any indications for surgical treatment of septal defects due to their low effect on the general hemodynamics.

Endovascular Procedure

The patient underwent combined endotracheal anesthesia with inhaled sevoflurane and intravenous fentanyl, propofol, and rocuronium. Due to a high risk of hemodynamic disorders and the need for controlled hypotension during the main intervention, invasive arterial monitoring was performed.

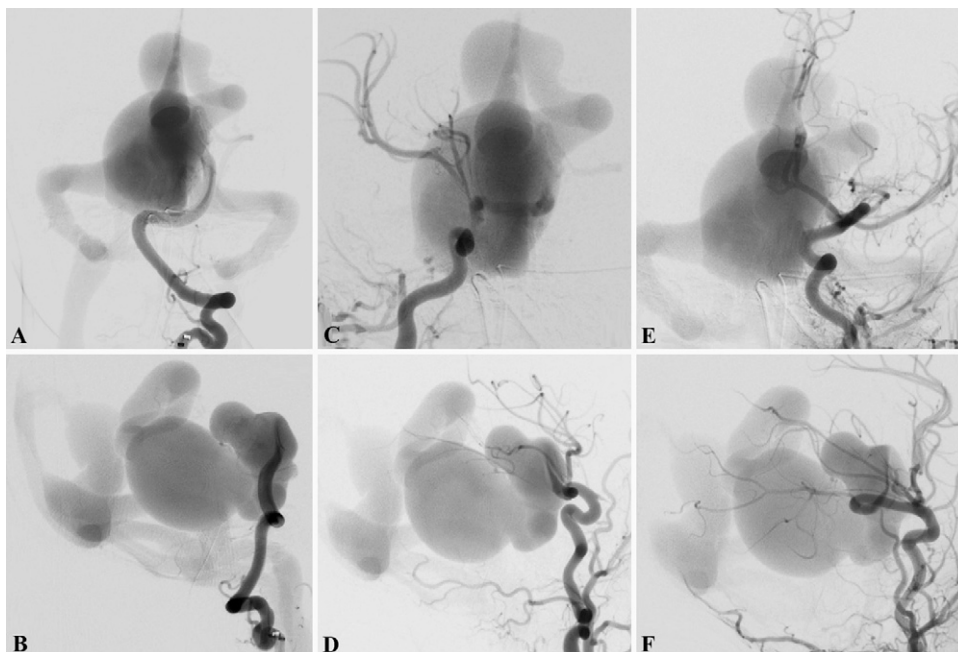


FIG. 2. Angioarchitectonics of the AVF on anteroposterior (**A, C, and E**) and lateral (**B, D, and F**) angiograms of the vertebral (**A and B**), right carotid (**C and D**), and left carotid (**E and F**) arteries.

A 5-French sheath was introduced into the right femoral artery using a conventional Seldinger technique to access the arterial system. A Sophia 5-French catheter (Microvention) was chosen as a guiding catheter due to its lumen size suitable for staged angiography and minimal invasiveness to the newborn's vessels.

Cerebral angiography showed a direct communication between the basilar artery and the deep cerebral vein as well as varicose changes in the vein (maximum dilation 34×27 mm and 23×14 mm). The blood further entered the vein of Galen and the straight sinus; all venous drainage routes were significantly dilated. The blood also flowed in the AVF from both carotid systems through dilated posterior communicating arteries (PCoAs) and P1 segments of both PCoAs originating from the basilar artery in the immediate vicinity of the AV shunt site (Fig. 2).

Detachable microcoils were used to close the AVF. For this, a PX SLIM microcatheter (Penumbra) was introduced through the fistula into the first dilation; six PC400 microcoils (Penumbra) with a coil diameter of 10–22 mm and a total length of 297 cm were placed in the vein (Fig. 3). This resulted in subtotal thrombosis of the fistula with a significant decrease in the AV shunting and a significant improvement in contrast enhancement of cerebral vessels.

Cerebral hemodynamics were intraoperatively monitored through the anterior fontanel using Doppler ultrasound. Before embolization, turbulent blood flow at a velocity of 110 cm/s was determined in the fistula. After embolization, a fourfold decrease in blood flow through

the fistula was observed (biphasic blood flow with a maximum velocity of 27 cm/s).

The intervention was completed without technical difficulties; cone beam computed tomography (XperCT, Philips) at the end of procedure showed no signs of intracranial hemorrhage. A total of 12 ml of a nonionic iodine-containing contrast agent was used during the procedure. The total fluoroscopy time was 10 minutes.

Postoperative Management

After intervention, the patient was transferred to the pediatric intensive care unit. In order to avoid early hemorrhagic complications, the patient underwent prolonged postoperative midazolam sedation for 12 hours. No inotropic support was required in the early postoperative period; no signs of pulmonary hypertension were observed. The pulmonary artery pressure decreased to subnormal values (<37 mm Hg). All these signs indicated a significant decrease in the pathological shunt to the pulmonary circulation. The indications for patient activation included the absence of severe neurological symptoms, stable hemodynamic parameters, adequate diuresis, and normal acid–base balance parameters with minimal respiratory support. The patient was extubated on day 3 and was discharged from the intensive care unit on day 6 after procedure.

Left ptosis and exotropia due to left oculomotor nerve palsy (the third pair) were detected in the newborn in the postoperative period after awakening. No pyramidal and brainstem disorders were diagnosed; no evidence for alternating syndromes was found.

Brainstem compression by a thrombosed deep cerebral vein was the most likely cause of postoperative deficiency.

MRI at 6 hours and 18 days after intervention showed neither ischemic nor hemorrhagic changes in the brain (Fig. 4). Progressive thrombosis of the fistula, deep cerebral vein draining the fistula, vein of Galen, and straight sinus was observed; blood flow in the superior sagittal sinus, confluence of sinuses, and both transverse sinuses was retained. There were no signs of cerebral venous insufficiency and cerebral venous infarction. A gradual decrease in the size of the main (largest) draining vein varix and partial recanalization of the straight sinus combined with gradual improvement of the neurological deficit (the eye began to open slightly; strabismus decreased) developed.

The patient and his mother were discharged from the department on day 18 after intervention. The control examination (MRI and neurological status) at 1.5 and 6 months showed blood flow absence through the fistula, continued regression of oculomotor disorders, and a satisfactory condition of the infant without physical and mental developmental delay.

Discussion

Pediatric intracranial AV shunting malformations are some of the most complicated issues of modern medicine. An important aspect of this issue is an extremely high risk of an unfavorable natural course of the disease (up to 90%).³ Another equally important aspect is an extremely high risk of surgical treatment of the disease, especially in the first days of life.

Observations

For 10 years, our team at the Meshalkin National Medical Research Center and Federal Center of Brain Research and Neurotechnologies has accumulated extensive experience in endovascular treatment of pediatric patients with congenital cerebrovascular pathology; we have

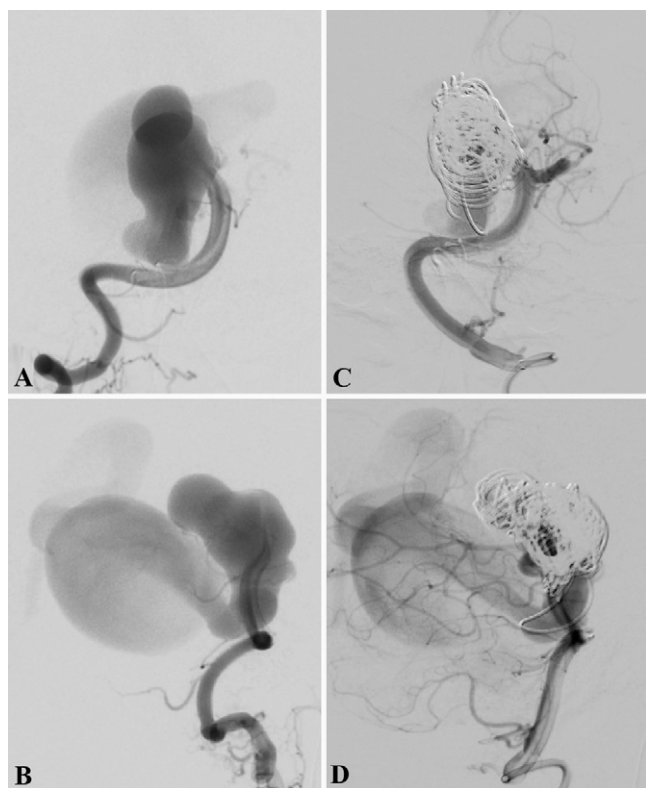


FIG. 3. Anteroposterior (A and C) and lateral (B and D) vertebral angiograms before (A and B) and after (C and D) AVF embolization in the same phase of contrast enhancement. A significant improvement in contrast enhancement of cerebral vessels and a pronounced decrease in blood flow through the fistula are observed.

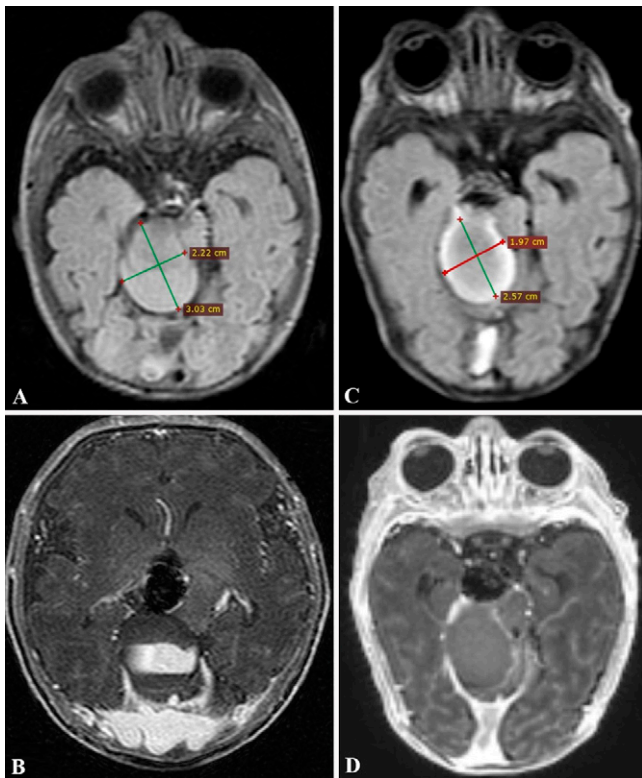


FIG. 4. Neonatal brain images obtained 6 hours (**A and B**) and 18 days (**C and D**) after intervention. Axial fluid-attenuated inversion recovery (**A and C**) and axial three-dimensional T1 turbo field echo (**B and D**) images show progressive thrombosis and some decrease in the size of the draining vein over time.

conducted a total of 158 surgeries in 70 patients, including 18 endovascular interventions in 16 newborns. In the presented case, the decision to perform fistula embolization in the first days after birth was made on the basis of the degree of hemodynamic load imposed by the fistula on the whole body. In cases of severe pulmonary hypertension and congestive heart failure with a high risk of death, intervention was a lifesaving and urgent procedure. We also used the Bicêtre neonatal evaluation score to assess the child's condition and determine indications for intervention. However, due to the difficulty of predicting long-term outcomes, it was not used as the main score when deciding whether to perform surgery. In the presented case, any delay might cause rapid decompensation and death.

On the basis of our experience of using various embolic agents, we believe that cyanoacrylate compositions are the most suitable for AVF embolization. Due to their adhesive properties, they allow controlled and effective occlusion of high-flow fistulas (>70% of procedures in our series of cases). Nonadhesive compositions are effective for final embolization of residual small feeders of a vein of Galen AVM.

Lessons

In the presented case, the use of nonadhesive agents was technically impossible due to high blood flow, whereas application of adhesive agents was associated with an extremely high risk of basilar artery occlusion and ischemia of the brainstem and subcortical nuclei.

Therefore, we decided to use subtotal embolization of the AVF with 20-inch soft detachable coils. Using detachable microcoils significantly reduced the AV shunt through the fistula with almost complete relief of symptoms of venous hypertension and overload of the right heart chambers, and it also prevented progressive reactive thrombosis of the major arteries. The use of microcoils and gradual thrombosis of the draining vein prevented enlargement of varicose veins and severe brainstem compression.⁶ Subsequent follow-up examinations showed a gradual lysis of thrombi in the draining vein, which correlated well with regression of oculomotor disorders. In the follow-up period, the child had no delay in growth and development.

In this paper, we report a rare case of a direct communication between the basilar artery and a large cerebral vein in a newborn. This condition is life threatening and requires urgent treatment. Wide availability of endovascular tools and experience of using them allows one to optimize the embolization strategy in each specific case. In the presented case, we used incomplete coil embolization of AVF, which provided a good clinical outcome.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Berestov, Seleznev, Kiselev, Brusyanskaya, Orlov. Acquisition of data: Berestov, Obedinskaya, Korostyshevskaya, Gofer, Bondarenko, Brusyanskaya. Analysis and interpretation of data: Berestov, Obedinskaya, Korostyshevskaya, Gofer, Kiselev, Brusyanskaya. Drafting the article: Berestov, Kiselev. Critically revising the article: Berestov, Kiselev, Krasilnikov, Orlov. Reviewed submitted version of manuscript: Berestov, Krasilnikov. Approved the final version of the manuscript on behalf of all authors: Berestov. Statistical analysis: Berestov. Administrative/technical/material support: Berestov, Orlov. Study supervision: Berestov, Orlov.

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