

A case of PAVF diagnosed soon after birth and given cerebrovascular therapy 4 months after birth is reported.

CASE DESCRIPTION: The patient presented with heart failure immediately after birth. Ultrasonography of the head showed abnormal blood flow in the brain. On digital subtraction angiography performed 4 months after birth, a PAVF with a dural feeder shunt and a giant varix at the posterior temporal part was confirmed. After transarterial embolization (TAE), shunt blood flow disappeared. New shunt flow from the right posterior cerebral artery into the varix was confirmed by magnetic resonance imaging 3 months after the operation. A second TAE procedure using a liquid embolic material was performed and confirmed the complete disappearance of the shunt.

CONCLUSIONS: This report describes a case of infant PAVF with heart failure, a giant varix, hydrocephalus, and intraventricular hemorrhage treated by TAE using platinum coils and liquid embolic material ¹⁾.

Chugh et al. present 2 pediatric patients who were both found to have pial arteriovenous fistulas (AVFs) with subsequent genetic analysis revealing mutations in the [RASA1](#) gene. Considering their family history of distinct cutaneous lesions, these mutations were likely inherited as opposed to de novo mutations. Patient 1 had large capillary malformations on the left side of the face and neck, associated with macrocephaly, and presented at the age of 32 months with speech delay, right-sided weakness, and focal seizures involving the right side of the body. Patient 2 presented with proptosis at the age of 9 months, but was otherwise neurologically intact. Given the chance for definitive single-stage control of vascular shunt (obviating chances for radiation exposure with endovascular treatment) and surgically accessible location of these intracranial lesions, both patients were treated with surgery with excellent clinical and radiological outcome. In general, given the high mortality secondary to severe congestive heart failure when treated conservatively, the goal of treatment in cortical AVF in young children, even when asymptomatic, is rapid control of the shunt. This was achieved successfully in our cases - both patients experienced significant symptomatic improvement following surgery and remained neurologically stable in the subsequent follow-up visits ²⁾.

2015

Lo Presti et al. present the case of a 15-year-old boy with [Sickle cell disease](#) SCD-associated moyamoya disease harboring a pAVF who developed a de novo venous aneurysm 8 months after undergoing indirect superficial temporal artery-middle cerebral artery (MCA) bypass that was complicated by bilateral ischemia of the MCA territory. The pAVF was successfully treated with transarterial embolization using Onyx. The authors describe the possible pathophysiological mechanisms and management strategies for this rare occurrence ³⁾.

2014

A rare case of pediatric pial AVF treated by direct disconnection with the aid of [indocyanine green videoangiography](#). A 3-year-old girl presented with [developmental problems](#). [Magnetic resonance imaging](#) revealed [brain atrophy](#) and an anomalous left temporal [vascular mass](#). [Angiography](#) showed a high-flow pial AVF in the early arterial phase fed by the [M1](#) portion of the left [middle cerebral artery](#) and draining into the [superficial sylvian vein](#) and the [vein of Trolard](#) with a large varix. Given that her fistula was located in a superficial region that was easily accessible by [craniotomy](#), the authors

successfully disconnected her pial AVF by direct surgery aided by ICG videoangiography, which clearly confirmed the shunting point ⁴⁾.

1)

Okazaki T, Sakamoto S, Ishii D, Oshita J, Matsushige T, Shinagawa K, Ichinose N, Matsuda S, Kurisu K. A Pial Arteriovenous Fistula in Infancy as the Presenting Manifestation of Hereditary Hemorrhagic Telangiectasia. *World Neurosurg.* 2019 Feb;122:322-325. doi: 10.1016/j.wneu.2018.10.146. Epub 2018 Nov 1. PubMed PMID: 30391600.

2)

Chugh AJ, Shahid A, Manjila S, Gulati D, Bambakidis NC. Pial Arteriovenous Fistula and Capillary Malformation-Arteriovenous Malformation Associated with RASA1 Mutation: 2 Pediatric Cases with Successful Surgical Management. *Pediatr Neurosurg.* 2017 May 31. doi: 10.1159/000474942. [Epub ahead of print] PubMed PMID: 28558378.

3)

Lo Presti A, Weil AG, Fallah A, Peterson EC, Niazi TN, Bhatia S. Treatment of a cerebral pial arteriovenous fistula in a patient with sickle cell disease-related moyamoya syndrome: case report. *J Neurosurg Pediatr.* 2015 May 22:1-5. [Epub ahead of print] PubMed PMID: 26053963.

4)

Sugimoto T, Park YS, Nakagawa I, Nishimura F, Motoyama Y, Nakase H. Effectiveness of intraoperative indocyanine green videoangiography in direct surgical treatment of pediatric intracranial pial arteriovenous fistula. *J Neurosurg Pediatr.* 2014 Oct 24:1-5. [Epub ahead of print] PubMed PMID: 25343732.

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