
Hemimegalencephaly and Intractable Epilepsy Treated with Embolic Hemispherectomy

John M. Mathis, John D. Barr, A. Leland Albright, and Joseph A. Horton

Summary: A patient with hemimegalencephaly and intractable epilepsy underwent a preoperative embolic hemispherectomy. A seizure-free interval of 1 year followed the embolization procedure. In addition, the procedure was thought to be beneficial in limiting blood loss during a subsequent surgical hemispherectomy.

Index terms: Megalencephaly; Seizures; Surgery, resective; Interventional neuroradiology

Hemimegalencephaly is a rare dysplastic malformation of the brain resulting from overgrowth of part or all of a cerebral hemisphere. Clinical findings include psychomotor retardation, hemiparesis, and intractable seizures. Early control of seizures is needed to allow maximal psychomotor development. Seizures associated with hemimegalencephaly are difficult or impossible to control medically. Hemispherectomy has been used in several series to effectively control seizures (1–9). This report describes a patient in whom staged cerebral embolization was used to achieve initial seizure control and was a preoperative adjunct to surgical hemispherectomy.

Case Report

A 2829-g girl was born after an uncomplicated 39-week gestation and delivery. The child was considered well when discharged home on the third day of life. She was readmitted at 17 days of age for evaluation of frequent seizure activity localized to the right cerebral hemisphere. Subsequent magnetic resonance (MR) imaging showed findings of hemimegalencephaly (Fig 1A and B). The right hemisphere was enlarged with thickened cortex and had an abnormal gray-white matter distribution. The right lateral ventricle also was enlarged.

Seizure frequency and duration increased, so that seizures occurred typically more than once per hour. Seizures were refractory to medical therapy including phenobarbital and adrenocorticotrophic hormone. Because of the intractable nature of the seizures, hemispherectomy was considered. It was suggested by the neurosurgical team that hemispherectomy be preceded by staged cerebral embolization to minimize operative blood loss and possibly decrease the postoperative risk of hydrocephalus and hemosiderosis. After extended conferences with the infant's parents, the first stage of embolization was undertaken at 10 weeks of age.

Embolization of the right anterior cerebral artery distribution was performed first, with the consideration that this relatively small segment of the hemisphere would create less swelling and mass effect than would the middle cerebral artery territory. The resulting encephalomalacia would then provide a margin of safety if more mass effect were experienced after embolization of the middle cerebral artery.

With the patient under general anesthesia, a coaxial system was introduced via the right common femoral artery with a 5F catheter as a base. A Tracker 18 catheter (Target Therapeutics, Fremont, Calif) was positioned beyond the anterior communicating artery in the right anterior cerebral artery (Fig 1C). Embolization was performed with a mixture of powdered absorbable gelatin sponge (Gelfoam) and 200 to 300 μm of polyvinyl alcohol particles suspended in nonionic contrast material. All branches of the anterior cerebral artery distal to the anterior communicating artery were embolized (Fig 1D). Computed tomography (CT) 24 hours after embolization showed infarction of the distribution of the anterior cerebral artery (Fig 1E). There was no significant alteration in seizures after the embolization.

Embolizations of cortical branches of the middle cerebral (Fig 1F and G) and posterior cerebral arteries were performed in a similar manner at 10-week intervals after embolization of the anterior cerebral artery. In each case, catheter placement was distal to perforating vessels (len-

Received August 18, 1993; accepted after revision February 25, 1994.

From the Divisions of Neuroradiology (J.M.M., J.D.B., J.A.H.) and Pediatric Neurosurgery (A.L.A.), University of Pittsburgh (Pa); the Division of Neuroradiology–Endovascular Therapy, University of Virginia Health Sciences Center, Charlottesville (J.M.M.); and the Division of Neuroradiology, Pennsylvania State University, Hershey (J.D.B.)

Address reprint requests to John M. Mathis, MD, Director, Diagnostic and Interventional Neuroradiology, University of Maryland Medical System, 22 S Greene St, Baltimore, MD 21201.

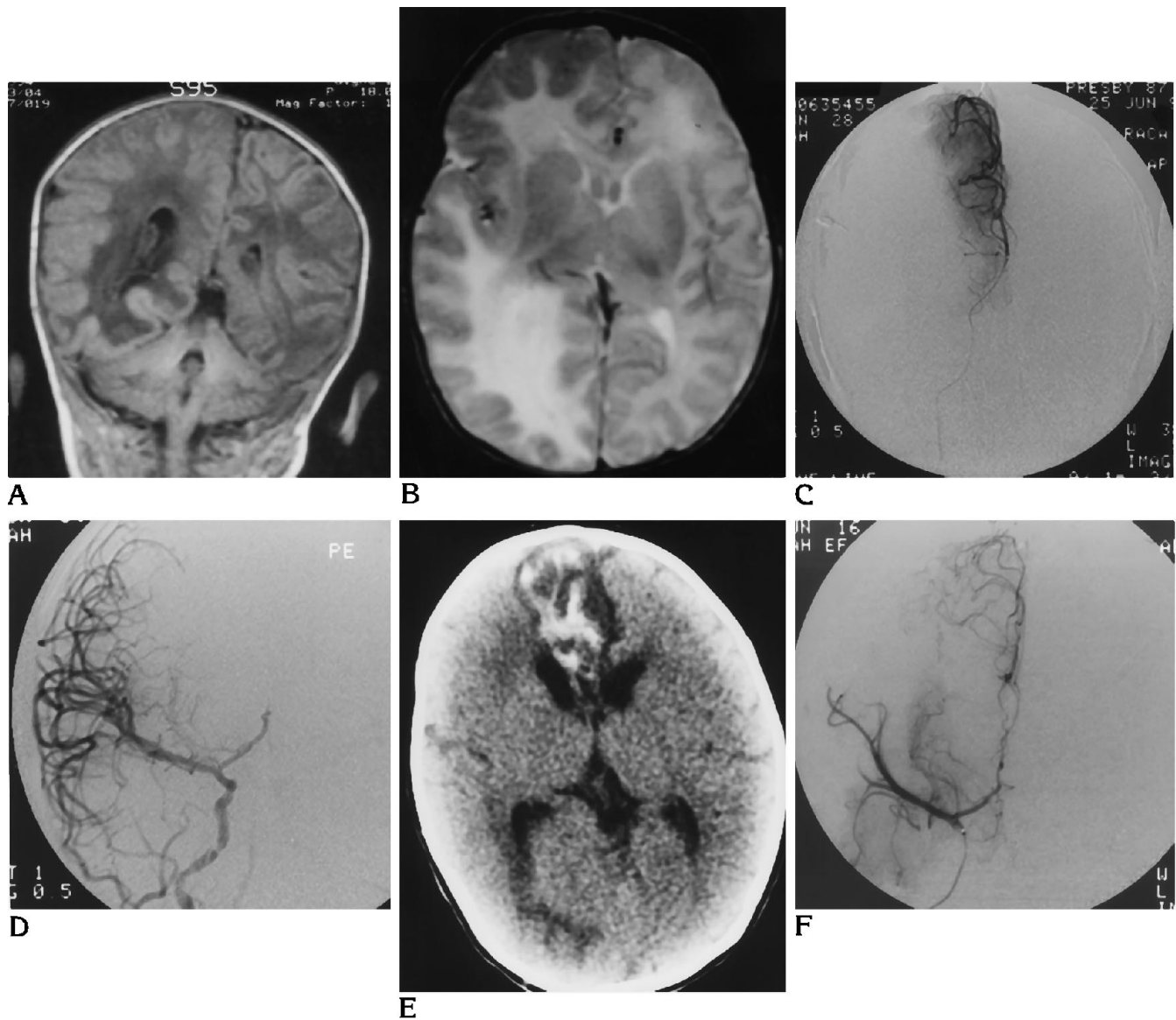


Fig 1. A, Coronal MR image with short repetition and echo times shows enlargement of the right hemisphere with right ventriculomegaly.

B, Axial MR image with long repetition and echo times also shows enlargement of the right hemisphere. Abnormal white matter signal and cortical dysplasia are present on the right as well.

C, Superselective preembolization injection of the territory of the right anterior cerebral artery with microcatheter positioned beyond the anterior communicating and perforating vessels.

D, Postembolization injection of the right internal carotid artery shows occlusion of branches of the right anterior cerebral artery beyond the anterior communicating level.

E, CT scan 24 hours after embolization of the right anterior cerebral artery shows high attenuation from the embolic agent and relatively mild mass effect.

F, Angiogram immediately after embolization of the cortical territory of the right middle cerebral artery. Recanalization is present in branches of the right anterior cerebral artery embolized 10 weeks earlier. (*Figure continues.*)

ticulostriate, thalamostriate, thalamoperforate, and thalamogeniculate). Provocative testing with amobarbital demonstrated loss of evoked potentials in the selected hemispheric regions before permanent embolization. The basal ganglia were intentionally spared. No complications occurred during embolization.

Seizure activity was present and relatively unchanged during the course of embolization. Approximately 2 weeks after the final (posterior cerebral artery) embolization, clinically evident seizures started to abate even though waves on the electroencephalogram remained epileptiform, particularly in the occipital region. At 6 and 10 months after

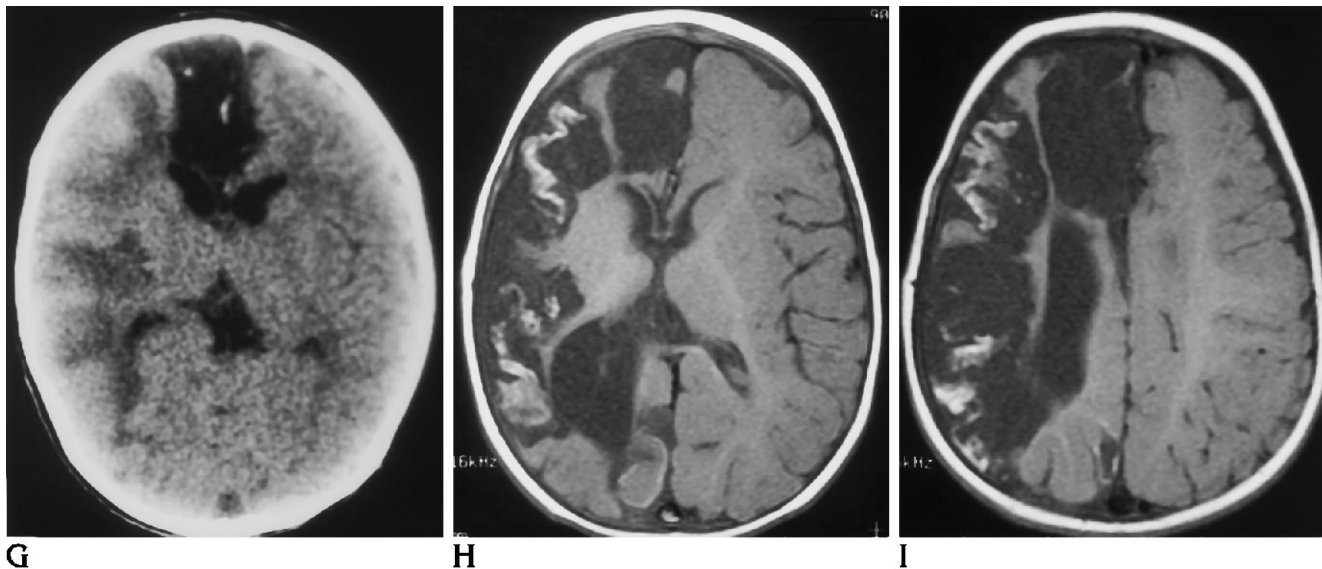


Fig 1, *cont'd.* G, CT scan 24 hours after embolization of the territory of the right middle cerebral artery shows local edema and high attenuation secondary to embolic material. Encephalomalacia is present in the anterior cerebral artery territory embolized 10 weeks earlier.

H and I, MR images (short repetition and echo times) 14 months after embolic hemispherectomy show encephalomalacia in the right anterior and middle cerebral artery territories with smaller changes in the region of the right posterior cerebral artery. Sparing of the basal ganglia is noted in I.

completion of embolization, the patient was free of clinical seizures on a small residual dose of phenobarbital. The left hemiparesis was abating; she could stand and was beginning to take steps but had no dexterity of the left fingers.

At 14 months after embolization, clinical seizure activity resumed and ultimately became intractable to maximal medical therapy. MR showed encephalomalacia in the territories of the anterior and middle cerebral arteries with sparing of basal ganglia, but little apparent change in the distribution of the posterior cerebral artery (Fig 1H and I). An electroencephalogram demonstrated considerable epileptiform activity in the right occipital region. The neurosurgical team elected to perform a hemispherectomy rather than attempt another embolization. At operation, electrocorticography revealed nearly continuous epileptiform activity in the parietooccipital region; the right temporal lobe and parietooccipital cortex were removed. Postresection electrocorticography revealed persistent, frequent sharp-wave activity in the remaining embolized tissue; the remainder of the hemisphere was removed. Embolization was thought to be a positive adjunctive procedure that transiently controlled the seizures and minimized intraoperative blood loss. The child recovered from the procedure and was again seizure free.

Discussion

Hemimegalencephaly is a rare dysplastic malformation of the brain that was reported by Sims (10) in 1835. Macroscopically, there is hemispheric enlargement, ipsilateral ventriculomegaly, and indistinct gray-white matter junctions (4, 5, 11, 12).

Gyral patterns range from mild polymicrogyria to complete lissencephaly and correspond to variable degrees of heterotopia (1). Light microscopy reveals an increased number and size of glial cells, foci of calcification in subcortical white matter, and marked neuronal enlargement. Increased DNA content has been found, suggesting heteroploidy (13, 14). In addition, Rosenthal fibers have been discovered in giant glial cells on electron micrographs (4). Barkovich and Chuang (1) suggest that an insult to the developing brain in the middle to late second trimester could account for both polymicrogyria and overgrowth of hemispheric white matter.

Both CT and MR show hemispheric enlargement, ipsilateral ventriculomegaly, and an abnormal gyral and white matter pattern. Affected white matter has low attenuation on CT and demonstrates abnormal high signal on T2-weighted MR images (1).

Hemimegalencephaly can be an isolated finding (as in our patient) or associated with extracranial manifestations including hemihypertrophy (15), hypomelanosis of Ito (16), linear sebaceous nevus of Jadassohn (16), epidermal nevus (17), Klippel-Trenaunay-Weber syndrome (18), and neurofibromatosis (19, 20).

Patients with hemimegalencephaly often have hemiparesis, hemianopia, and variable

degrees of psychomotor retardation. However, intractable seizures often dominate the clinical presentation. Because seizures are usually refractory to medical therapy, hemispherectomy has been described by several authors as an effective method of seizure control (1-9, 21). If seizures are controlled early, psychomotor development can again progress, particularly if the basal ganglia are spared.

Before any type of invasive therapy, surgical or embolic, every effort must be made to establish seizure control medically. Even with a known lesion, long-term, continuous electroencephalographic monitoring is also needed to exclude an abnormal focus from a remote location. Embolization as a primary therapy for seizure control has not been previously described. Embolization in the present case and its association with a period when this patient was free from clinical seizures should not be construed as evidence of a beneficial causal effect. In the present case, it was requested as an adjunct to surgical hemispherectomy in an attempt to limit blood loss. As with hemispherectomy, embolization should spare diencephalic and basal ganglion structures. This can be accomplished by careful catheter placement beyond proximal perforating vessels, with embolization terminated once major vascular stasis occurs and before there is reflux.

Particle embolization has the potential disadvantage of delayed vascular recanalization. Acrylic embolization might offer an advantage if significant delay between embolization and surgery is planned. Staged embolizations were elected in this case to minimize complications that might be associated with ischemic mass effect. Even with this consideration, a more efficient approach could accomplish embolization in two stages, combining the anterior and posterior cerebral artery territories followed later by embolization of the distribution of the middle cerebral artery.

Embolic hemispherectomy was considered a helpful preoperative adjunct to surgical hemispherectomy in this patient. Initial seizure control after embolic hemispherectomy, though encouraging, cannot at this point be suggested as having a primary therapeutic role. At present, this technique is experimental and further investigation is needed.

References

1. Barkovich J, Chuang S. Unilateral megalencephaly: correlation of MR imaging and pathologic characteristics. *AJNR Am J Neuroradiol* 1990;11:523-531
2. Fitz C, Harwood-Nash D, Boldt D. The radiographic features of unilateral megalencephaly. *Neuroradiology* 1978;16:145-148
3. Fusco L, Vigeveno F. Reversible operculum syndrome caused by progressive epilepsy partialis continua in a child with left hemimegalencephaly. *J Neurol Neurosurg Psychiatry* 1991;54:556-558
4. Robain O, Chiron C, Dulac O. Electron microscopic and Golgi study in a case of hemispherectomy performed because of hemimegalencephaly. *Acta Neuropathol (Berl)* 1989;77:664-666
5. Trounce J, Rutter N, Mellor D. Hemimegalencephaly: diagnosis and treatment. *Dev Med Child Neurol* 1991;33:261-266
6. Vigeveno F, Bertini E, Boldrini R, et al. Hemimegalencephaly and intractable epilepsy: benefits of hemispherectomy. *Epilepsia* 1989;30:833-843
7. Vigeveno F, di Rocco C. Effectiveness of hemispherectomy in hemimegalencephaly with intractable seizures. *Neuropediatrics* 1990;21:222-223
8. King M, Stephenson J. Hemimegalencephaly: a case for hemispherectomy? *Neuropediatrics* 1985;16:46-55
9. Towbin R, Witte D, Ball W, Han B. Pediatric case of the day: hemimegalencephaly. *Radiographics* 1988;8:573-577
10. Sims J. On hypertrophy and atrophy of the brain. *Medicochirurgical Transactions. R Med Chir Soc (London)* 1835;19:315-380
11. Tjiam A, Stefanko S, Schenk V, de Vlieger M. Infantile spasms associated with hemiphysarrhythmia and hemimegalencephaly. *Dev Med Child Neurol* 1978;20:779-789
12. Townsend J, Nielsen S, Malamud N. Unilateral megalencephaly: hamartoma or neoplasm. *Neurology* 1975;25:448-453
13. Bignami A, Palladini G, Zapella M. Unilateral megalencephaly with nerve cell hypertrophy: an anatomical and quantitative histochemical study. *Brain Res* 1968;9:103-114
14. Manz H, Phillips T, Rowden G, McCullough D. Unilateral megalencephaly, cerebral cortical dysplasia, neuronal hypertrophy and heterotopia. *Acta Neuropathol (Berl)* 1979;45:97-103
15. Cheruy M, Heller F. An unusual variant of Klippel-Trenaunay syndrome: association of total hemihypertrophy, hemimegalencephaly and bilateral extremity enlargement. *Acta Chir Belg* 1987;87:73-76
16. Peserico A, Battistella P, Bertoli P, Drigo P. Unilateral hypomelanosis of Ito with hemimegalencephaly. *Acta Paediatr* 1988;77:446-447
17. Sakuta R, Aikawa H, Takashima S, Yoza A, Ryo S. Epidermal nevus syndrome with hemimegalencephaly. *Brain Dev* 1989;11:191-194
18. Burke J, West N, Strachan I. Congenital nystagmus, anisomyopia and hemimegalencephaly in Klippel-Trenaunay-Weber syndrome. *J Pediatr Ophthalmol Strabismus* 1991;28:41-44
19. Cusmai R, Curatolo P, Mangano S, Cheminal R, Echenne B. Hemimegalencephaly and neurofibromatosis. *Neuropediatrics* 1990;21:179-182
20. Ross G, Miller J, Persing J, Ulrich H. Hemimegalencephaly, hemifacial hypertrophy and intracranial lipoma: a variant of neurofibromatosis. *Neurofibromatosis* 1989;2:69-77
21. Davis KG, Maxwell RE, French LA. Hemispherectomy for intractable seizures: long-term results in 17 patients followed for up to 38 years. *J Neurosurg* 1993;78:733-740