COMMENT. Moyamoya disease is a largely Korean or Japanese cerebrovascular disease of internal carotid arteries with compensatory vascular network at the base of the brain. Repeated transient ischemic attacks are common presenting symptoms in children and intracranial hemorrhage in adults. Surgery to establish adequate collateral circulation is effective in ischemic cases without infarction. Outcome was favorable in 81% of the above large series of pediatric cases. Infarction at the time of diagnosis of MMD is predictive of a 3-fold increased risk of unfavorable surgical outcome, whereas decreased vascular reserve only with normal basal perfusion on SPECT has a 14-fold increased frequency of favorable outcome. Long-term follow-up is recommended in unilateral cases to monitor potential progression and bilateral involvement.

DIAGNOSIS AND TREATMENT OF VEIN OF GALEN ANEURYSMAL MALFORMATIONS

Diagnosis and treatment of 13 patients with vein of Galen aneurysmal malformation (VGAM), seen at Children's Hospital and University of Pennsylvania, Philadelphia, 1994-2007, were evaluated retrospectively, and those treated within the first 2 weeks were compared with those treated later. Six (46%) patients were diagnosed in utero by fetal ultrasound or fetal MR, three within 1 week of birth with cardiac failure, and four from 1.5 to 31 months of age. Cardiac failure was the most common presenting symptom, diagnosed in the first week of birth, and was present in 7 patients. Increasing head circumference, hydrocephalus and developmental delay characterized all but one of the patients presenting outside the neonatal period. One patient presented with subarachnoid hemorrhage at 31 months of age. Postnatal MRAs demonstrated preembolization hemorrhage in 4 patients, ischemia in 4, and diffuse parenchymal abnormality in 5. Five had hydrocephalus. Angioarchitecture of a choroidal pattern was demonstrated in 62% of VGAMs, and in all VGAMs presenting early with cardiac failure. Five of 6 presenting later in life had mural angioarchitecture. Endovascular treatment was implemented in 11 patients. Of 6 patients treated emergently in the first 2 weeks, 2 developed normally, 1 had mild to moderate neurological deficits, 1 had severe deficits, and 2 died. Outcome in 5 patients treated later, at 1.5 to 31 months, was better, normal in 3 and mild neurological deficits in 2. (Heuer GG, Gabel B, Beslow LA, et al. Diagnosis and treatment of vein of Galen aneurysmal malformations. **Childs Nerv Syst** July 2010;26:879-887). (Respond: Dr RW Hurst, Division of Neuroradiology, Hospital of the University of Pennsylvania, 3400 Spruce Street, Philadelphia, PA 19104. E-mail: Robert.Hurst@uphs.upenn.edu).

COMMENT. Cardiac failure is the presenting feature of neonatal cases of VGAM, and enlarging head circumference, hydrocephalus and developmental delay characterize cases with later onset. Endovascular management is the preferred approach in all age groups. Outcome in cases presenting after the neonatal period is better than those of early onset. Cardiac failure is particularly intractable, with mortality rates as high as 62%. In utero MR diagnosis allows for early stabilization and cardiac management.