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Aneurysmal malformation of the vein of Galen in three patients: clinical and radiological follow-up

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Abstract We studied three patients with aneurysmal malformations of the vein of Galen: all underwent embolisation followed by MRI and conventional angiography; two also underwent postembolisation magnetic resonance angiography (MRA). MRI was performed before treatment in two patients, MRA in only one, diagnosed prenatally. Two patients had normal appearances on follow-up MR studies and were completely cured by embolisation. However, the last patient, after two embolisations, despite the stabilisation of the clinical condition, underwent surgery and died. We think MRI is mandatory before endoarterial treatment, to assess the conditions of the brain. Angiography is mandatory only at the time of endovascular treatment, while MRA and MRI have a role in follow-up. Endoarterial embolisation remains the treatment of choice, and surgery is not advisable.

Key words Vein of Galen aneurysmal malformation · Magnetic resonance imaging · Angiography · Treatment

Introduction

Vein of Galen aneurysmal malformations (VGAM) are rare [1]. They are true congenital arteriovenous lesions [2] and need optimal early care. We describe three embolised patients assessed by serial MRI, and magnetic resonance (MRA) and conventional angiography.

Case reports

Case 1

A 3 kg newborn boy presented with mild heart failure which spontaneously regressed during the first month. MRI at 4 months demonstrated a mural VGAM with normal brain parenchyma (Fig. 1 a). The child underwent two embolisations when he was 7 and 10 months old, with only partial occlusion of the malformation. Postembolisation MRI and MRA at 16, 28 and 38 months showed the glue cast in the VGAM and progressive further occlusion (Fig. 1 b, c). Conventional angiography at 20 and 39 months (Fig. 1 d) confirmed occlusion of the VGAM. At present the patient's neurological and cognitive status is normal.

Case 2

An 8-month-old boy presented with increased head circumference, moderate cardiac failure, epileptic seizures and mild developmental delay. CT showed a large VGAM, and enlargement of the third and lateral ventricles; no focal brain lesion was observed. An internal carotid angiogram confirmed the presence of the VGAM (Fig. 2 a), which was completely embolised at 10 and 12 months. Two weeks after the last embolisation, MRI showed thrombosis of the lesion; ventriculomegaly and macrocrania were still evident (Fig. 2 b). However, since he was clinically normal, the child did not undergo shunting. MRI at 26 and 37 months showed progressive resolution of the VGAM and spontaneous regression of the ventricle enlargement (Fig. 2 c). Conventional angiography during follow-up confirmed complete cure of the VGAM (Fig. 2 d). At present, the patient's neurological and cognitive function is normal.

Case 3

A VGAM was diagnosed during the 32nd week of pregnancy by Doppler ultrasonography, and studied at the 36th week by MRI, which showed a VGAM without thrombosis of the venous sinuses

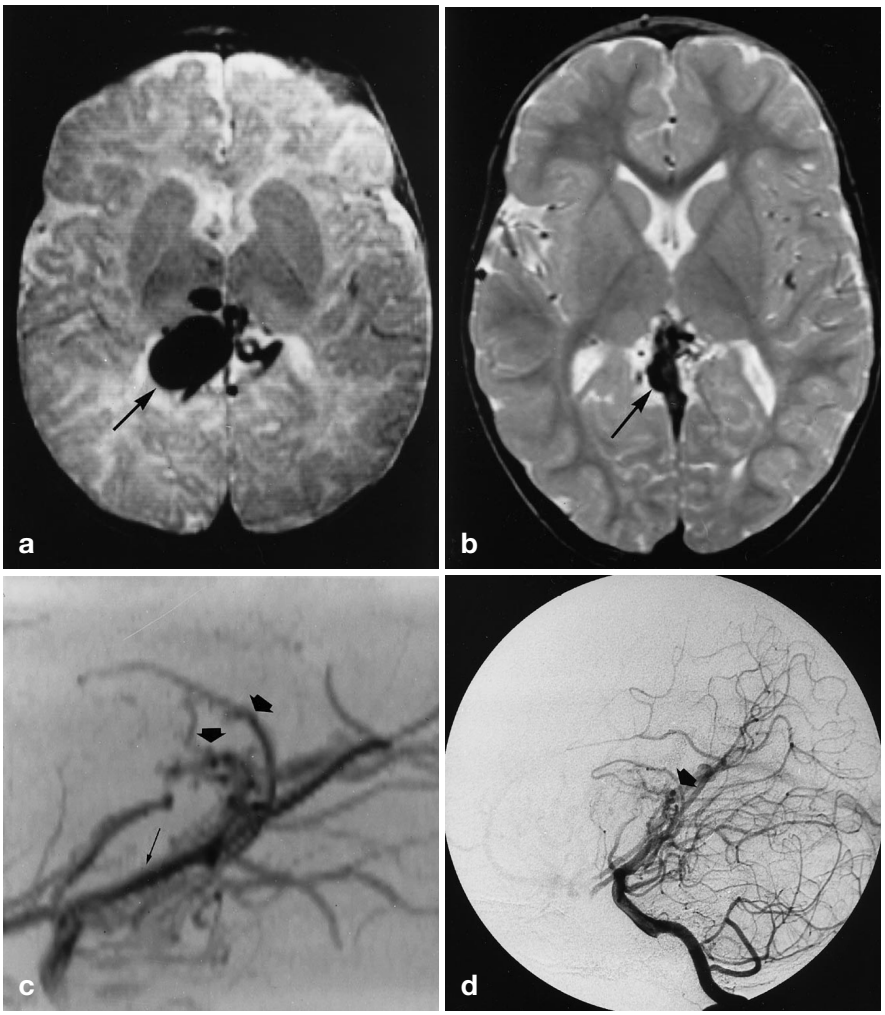

Fig. 1 Patient 1. VGAM diagnosed at 4 months of age. **a** Axial T2-weighted image at 4 months, showing malformation. No parenchymal brain abnormality. Normal size of ventricle. **b** Image at 38 months. Normal brain parenchyma and normal ventricle size. Mild dilatation of vein of Galen *(arrow)*. **c** 3D MRA: occlusion of the VGAM. Persistent enlargement of posterior choroidal *(large arrows)* and cerebral *(arrow)* arteries. **d** Follow-up left vertebral angiogram at 39 months: occlusion of the VGAM, with a very small shunt via a choroidal artery *(arrow)*

or any other arteriovenous malformation [3]. The brain parenchyma was normal, with slight enlargement of the right lateral ventricle; there was also a mild dilatation of the heart. The baby girl was born at 39 weeks. She was healthy except for mild, transient heart failure. At 2 months of age, she underwent MRI (Fig. 3a) which showed the VGAM better. It had moderate mass effect on the third ventricle and hypothalamus, with early dilatation of the lateral ventricles; the head circumference was normal. There were no focal brain lesions and the cerebellar tonsils were in normal position. MRA demonstrated the VGAM and dilated posterior choroidal, cerebral and basilar arteries. At the end of the third month, there was delay in growth and hydrodynamic problems, with progressive increase of the head and dilatation of the third and lateral ventricles. Because of these findings, early embolisation was planned.

This was performed at 4 months with N-butyl-cyanoacrylate (NBCA). After a first procedure, the baby was clinically well: her growth improved and the macrocrania did not progress. At 6 months, MRI and MRA showed incomplete occlusion of the VGAM with residual slow flow and a small anterior portion still evident on the left (Fig. 3b). Despite further ventricular enlargement, the child's clinical condition was good. At 8 months, a second embolisation with NBCA occluded the malformation, but was complicated by an occipital hematoma. The patient developed severe intracranial hypertension and underwent ventricular shunting. One month later, she had recovered and neurological examination was normal. At 13 months after birth, MRI showed a collapsed, shunted left lateral ventricle and a small occipital subdural haematoma (Fig. 3c). On MRA, the VGAM seemed to be only very slightly patent (Fig. 3d) and the patient's clinical condition contraindicated further immediate embolisation. Nevertheless, the referring neurosurgeon (in a different institution) decided to approach the VGAM directly; unfortunately there were severe haemorrhagic complications, which led to the death of the patient.

Discussion

The VGAM is a true arteriovenous malformation occurring at the end of the embryonic period [1, 2]. It represents a dilatation of the persistent embryonic median prosencephalic vein of Markowski, with direct arteriovenous shunts [3]. On angioarchitectural analysis,

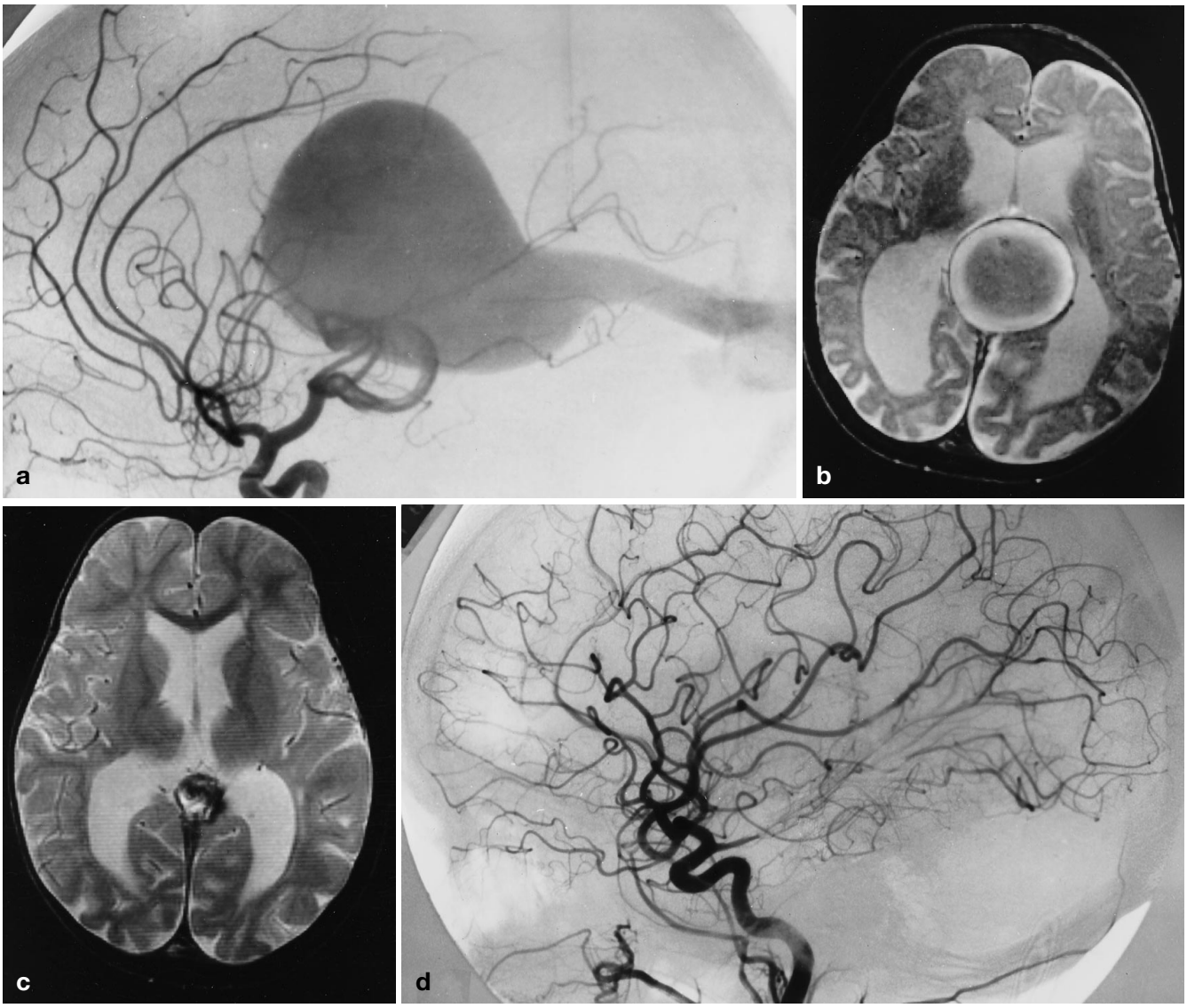

Fig. 2 Patient 2. VGAM diagnosed at 8 months of age. **a** Pre-embolisation right internal carotid angiogram at 10 months, confirming a mural VGAM. **b** Axial T2-weighted image 2 weeks after embolisation at 12 months. Normal brain parenchyma, but enlargement of the lateral ventricles persists. **c** Image following embolisation at 37 months. Complete occlusion and regression of the VGAM. Lateral ventricles are smaller; normal brain parenchyma. **d** Follow-up internal carotid angiogram: complete occlusion of the VGAM

two types can be differentiated [1]: "mural", with arteriovenous shunts in the wall of the dilated vein and mild neurological deficits; and "choroid", in the choroid fissure with multiple shunts communicating secondarily with the dilated median prosencephalic vein.

Several groups have contributed significantly to the understanding and management of VGAM [1–12], and the natural history of this disease is now well known [1, 2, 4]. The clinical features may differ according to the patient's age: in the neonate, there is early heart or multiorgan failure; in the infant, there are hydrodynamic disturbances, with macrocrania, hydrocephalus and a poor prognosis if venous thrombosis occurs; in childhood, developmental delay; and, in late childhood, epileptic seizures related to chronic venous ischaemia and neurological deficits, can occur.

The cause of hydrocephalus in these patients is debated. The controversy is whether venous hypertension and alterations in cerebrospinal fluid (CSF) resorption or obstruction of the ventricular system cause hydrocephalus. In our patient 2, hydrocephalus seemed to be related to obstruction of the ventricular system due to compression of the aqueduct of Sylvius by the VGAM; the fourth ventricle and extra-axial fluid spaces were

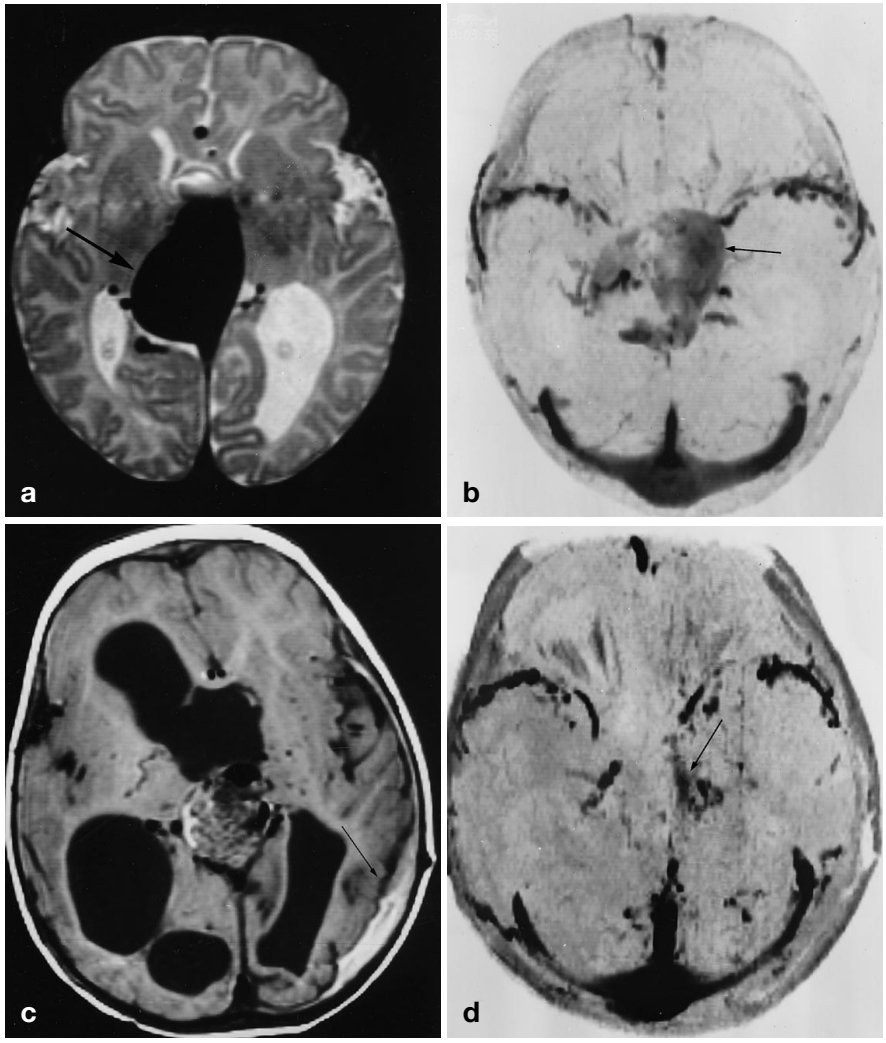

Fig. 3 Patient 3. VGAM diagnosed antenatally. **a** Axial T2-weighted image at 2 months of age. The diagnosis of VGAM *(arrow)* is confirmed. Mild dilatation of the third and lateral ventricles, without focal brain lesions. **b** Axial 2D MRA postembolisation, at 6 months: incomplete occlusion of VGAM, a small anterior-left portion *(arrow)* remaining. **c** Axial T1-weighted image at 13 months after ventricular shunting: small left occipital subdural haematoma *(arrow)* and collapsed lateral ventricle. **d** Axial 2D MRA at 13 months: further occlusion of the VGAM with only a very small portion still patent *(arrow)*

only minimally prominent. CSF flow studies would be helpful for determining the patency of the aqueduct, which may have had particular implications in this case, but unfortunately this technique was not available on our MRI system. Resolution of the aneurysmal dilatation of the vein of Galen and compression of the midbrain is observed with resolution of the hydrocephalus, as is a decrease in venous pressure. In patient 3, on the other hand, venous hypertension and alterations in CSF resorption appear to have played an important role in the development of hydrocephalus.

Some workers suggest that immaturity of the systems for absorption of CSF is the dominant factor in the development of the hydrodynamic disorders in infants [2, 6]. At this age, the venous system is responsible for the venous drainage of the brain and for absorption of extracellular brain water. Neither arachnoid granulations nor cavernous sinuses are functional at this stage and the function of the former may be delayed by venous hypertension. In patients with VGAM, the venous network becomes a high pressure system and, therefore, the resorption gradient between ventricles, extracellular space, medullary veins and dural sinuses easily fails. Macrocrania is the first sign of this imbalance; it progresses to ventriculomegaly and, when the sutures close, to subependymal atrophy. The main clinical consequence is developmental delay. Treatment consists in restoring physiological conditions by reducing venous hypertension. Ventricular shunting suddenly inverts the pressure gradient without treating the causes of the hydrodynamic disorders [6]. Shunting may therefore exacerbate the clinical deficits [6, 13], as in our patient 3, who was shunted after almost complete occlusion of the malformation. In patient 2 the ventriculomegaly regressed after embolisation, without any shunting procedure. Abstention from surgery is often advisable. In patients with thrombosed vein of Galen aneurysms, attempts to resect the occluded an-

eurysm sacs have been associated with unacceptable morbidity [11, 14].

Some MRI studies support the concept that hydrodynamic disorders, common in infants with VGAM [6], do not seem to be due to compression of the aqueduct. They support pathophysiological studies [15–18], which suggest an important role for increased venous pressure. Conventional angiography remains the method of choice for studying VGAM and in the majority of previous studies, was the only technique used [19, 20]. However, conventional angiography should not be the usual *diagnostic* procedure. Ultrasonography (US) is the modality of choice for investigation of neonatal abnormalities, because of its accuracy and safety. CT in patient 2 appeared more than adequate for diagnosis, but did not detect possible parenchymal damage. The benefits of MRI include providing data on the soft tissues, excellent soft-tissue contrast, and lack of interference by bony structures; MRI is mandatory for accurate assessment of the brain parenchyma. In VGAM it has a high prognostic value, allowing the decision about which therapeutic approach should be used and when. If there is severe parenchymal damage, endovascular treatment cannot compensate for the irreversible melting-brain process. MRI can help in the diagnosis particularly if this is antenatal [5, 7], and in understanding the pathophysiology of the disease [21]. It can be used to document the disproportionate growth of the cranial vault in comparison to the base. This can occur during the natural history of the VGAM, when there are long-standing hydrodynamic disorders, with thrombosis of the sigmoid sinus and of jugular veins. This condition provokes congestion of the posterior cranial fossa veins, and since there are no outlets and the posterior cranial fossa is small, without fontanelles the cerebellar tonsils can prolapse. MRI can also demonstrate the collapse of ventricles and the reversibility of cerebellar tonsillar prolapse after embolisation [6, 21]. MRI rarely shows the disappearance of the VGAM after spontaneous thrombosis [22]. However there are no serial MR studies on post-treatment follow-up: the published series are usually very small [20, 23, 24] or have only CT and angiographic data [25].

Conventional angiography need be carried out only if embolisation is planned, and at least once after the last embolisation. We recommend MRI and MRA during the pre-treatment period at 3 and 6 months after birth, then, following treatment, if clinical conditions are stable, at 6, 12, 24 and 36 months after the last treatment. Unfortunately, MRA, does not at present allow appropriate assessment of remodelling of veins, always observed in patients with VGAM after a cure. However, direct comparison of MRA and conventional angiography should be made in more patients, so that the multiplanar capabilities of MRA obviate serial angiography in these cases.

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References

1. Berenstein A, Lasjaunias P (1992) Surgical neuroangiography, Vol 4. Springer Verlag, Berlin Heidelberg New York, pp 267–317
2. Lasjaunias P (1997) Vascular diseases in neonates, infants and children. Springer Verlag, Berlin Heidelberg, pp 67–202
3. Raybaud CA, Strother CM, Hald JK (1989) Aneurysm of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. Neuroradiology 31: 109–128
4. Lasjaunias PL, Alvarez H, Rodesch G, García-Mónaco R, Terbrugge K, Burrows P, Taylor W (1996) Aneurysmal malformations of the vein of Galen. Follow-up of 120 children treated between 1984 and 1994. Intervent Neuroradiol 2: 15–26
5. Rodesch G, Hui F, Alvarez H, Tanaka A, Lasjaunias P (1994) Prognosis of antenatally diagnosed vein of Galen aneurysmal malformations. Child's Nerv Syst 10: 79–83
6. Zerah M, García-Mónaco R (1992) Hydrodynamics in vein of Galen malformation in 43 cases. Child's Nerv Syst 8: 111–117
7. Campi A, Scotti G, Filippi M, Gerevini S, Strigimi F, Lasjaunias P (1996) Antenatal diagnosis of vein of Galen aneurysmal malformation: MR study of fetal brain and postnatal follow-up. Neuroradiology 38: 87–90
8. García-Mónaco R, De Victor D, Mann C, Annedouche A, Terbrugge K, Lasjaunias P (1991) Congestive cardiac manifestations of cerebrocranial arteriovenous shunts. Endovascular management in 30 children. Child's Nerv Syst 7: 48–52
9. Seidenwurm D, Berenstein A, Hyman A, Kowalski H (1991) Vein of Galen malformation: correlation of clinical presentation, arteriography, and MR imaging. AJNR 12: 347–354
10. Johnston IH, Whittle IR, Besser M, Morgan MK (1987) Vein of Galen malformations: diagnosis and management. Neurosurgery 20: 747–758
11. Yasargil MG, Antic J, Laciera R, Jain KK, Boone SC (1976) Arteriovenous malformations of the vein of Galen. Microsurgical treatment. Surg Neurol 16: 195–200
12. Wisoff JH, Berenstein A, Choi IS (1990) Management of vein of Galen malformations. Concepts Pediatr Neurosurg 10: 137–155
13. Schneider SJ, Wisoff JS, Epstein FJ (1992) Complications of ventriculoperitoneal shunt procedures in hydrocephalus associated with vein of Galen malformations in childhood. Neurosurgery 30: 706–708
14. Weir BK, Allen PB, Miller JD (1968) Excision of thrombosed vein of Galen aneurysm in an infant. Case report. J Neurosurg 29: 619–622
15. Johnston IH (1973) Reduced CSF absorption syndrome. Reappraisal of benign intracranial hypertension and related conditions. Lancet ii 418–420

16. Chazal J, Tanguy A, Massini B, Janny P, Vanneuville G (1985) Dilatation of the subarachnoid pericerebral spaces and absorption of cerebrospinal fluid in the infant. Anat Clin 1: 61–66
17. Janny P, Colnet G, Veyre A, Chazal J, Barretto LC (1981) Hydrocéphalie à pression normal. Etude pré- et post-opératoire de 56 cas. Neurochirurgie 27: 86–96
18. Epstein CM (1974) The distribution of intracranial forces in acute and chronic hydrocephalus. J Neurol Sci 21: 171–180
19. Lasjaunias P, Terbrugge K, López Ibor L, Chiu M, Flodmark O, Chuang S, Goasguen J (1987) The role of dural anomalies in vein of Galen aneurysms: report of six cases and review of the literature. AJNR 8: 185–192
20. Dowd CF, Halbach VV, Barnwell SL, Higashida RT, Edwards MS, Hieshima GB (1990) Transfemoral venous embolization of vein of Galen malformations. AJNR 11: 643–648
21. Girard N, Lasjaunias P, Taylor W (1994) Reversible tonsillar prolapse in vein of Galen aneurysmal malformations: report of eight cases and pathophysiological hypothesis. Child's Nerv Syst 10: 141–147
22. Kuroki K, Uozumi T, Arita K, Takechi A, Matsuura R, Fujidaka (1995) Spontaneous disappearance of an aneurysmal malformation of the vein of Galen. Neuroradiology 37: 244–246
23. Ciricillo SF, Edwards MSB, Schmidt KG, Hieshima GB (1990) Interventional neuroradiological management of vein of Galen in the neonate. Neurosurgery 27: 22–28
24. Casasco A, Lylyk P, Hodes JE, Kohan G, Aymard A, Merland JJ (1991) Percutaneous transvenous catheterization and embolization of vein of Galen aneurysms. Neurosurgery 28: 260–265
25. Martelli A, Scotti G, Harwood-Nash DC, Fitz CR, Chuang S (1980) Aneurysms of the vein of Galen in children: CT and angiographic correlation. Neuroradiology 20: 123–133