Endovascular remodeling technique for vein of Galen aneurysmal malformations—angiographic confirmation of a connection between the median prosencephalic vein and the deep venous system

Report of 2 cases

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✓ It is commonly believed that in vein of Galen aneurysmal malformations (VGAMs) venous structures normally constituting the deep or Galenic venous system, such as the internal cerebral vein (ICV) and the basal vein of Rosenthal, are not connected to the vein of Galen. In this report, the authors describe 2 cases of successfully treated VGAM in which drainage of an ICV into the vein of Galen was confirmed by follow-up angiography. Two mural types of VGAM were treated using transarterial glue embolization when 1 child was 5 months and the other was 6 months old. The postoperative outcomes for these babies were complete cures. Follow-up digital subtraction angiography obtained after 12 months (Case 1) and 6 months (Case 2) confirmed that the shrunken median prosencephalic vein connects with the deep venous system. The possibility of normal deep Galenic venous drainage must be considered in endovascular management of VGAM. The goal of endovascular intervention is to close only the ventral component of the dilated median prosencephalic vein. (DOI: 10.3171/PED-08/01/075)

KEY WORDS • internal cerebral vein • median prosencephalic vein • pediatric neurosurgery • transarterial embolization • vein of Galen aneurysmal malformation

Veinf of Galen aneurysmal malformation is a rare anomaly that has recently become detectable in utero on ultrasonography or prenatal MR imaging, but usually presents in neonates.1,2 Aggressive therapy is required to avoid or minimize early systemic complications as well as early and late nervous system complications. In recent years the survival rate has increased, due to improvement in endovascular treatments for this abnormality.3,6,8 We report on the angiographic confirmation that the dorsal component of the median prosencephalic vein connects with the deep venous system after endovascular intervention. Therapeutic management and embryonic anatomical analysis are included in this report on VGAMs.

Abbreviations used in this paper: AVF = arteriovenous fistula; ICA = internal carotid artery; ICV = internal cerebral vein; MR = magnetic resonance; PChA = posterior choroidal artery; VGAM = vein of Galen aneurysmal malformation.

Case Reports

Case 1

History and Examination. This child, who was the second born to her family, was found to harbor an abnormal supratentorial cystic lesion on a prenatal ultrasonography study performed at 36 weeks of gestation in the course of a normal pregnancy. At 41 weeks, a baby girl with a birth weight of 3388 g was delivered vaginally. The baby was referred for examination at 2 weeks of age, at which time her head circumference was 34.2 cm. A plain chest x-ray film revealed cardiomegaly, the cardiothoracic ratio was 74%, and an intracranial bruit was detected on examination. Magnetic resonance imaging demonstrated a dilated vascular structure corresponding to the vein of Galen draining into the straight sinus. The cardiomegaly responded quickly to digoxin and diuretic therapy, and the cardiothoracic ratio improved to 53%. Cerebral angiography performed when the patient was
4 months old revealed a direct single-vessel AVF to the vein of Galen that was fed by a long circumferential artery arising from the PChAs (Fig. 1A). There was no evidence of venous reflux from the vein of Galen into the deep veins (Fig. 1B). The venous pouch enlarged gradually, and congestive liver and renal dysfunction appeared at 5 months.

Operation. At 6 months of age, the child was treated via an endovascular approach. Following left vertebral artery catheterization, the right medial PChA was superselectively catheterized and embolized with 0.5 ml of glue. The embolic material was visualized in the feeding vessel and partially in the ventral component of the Galenic pouch. Immediate follow-up angiography demonstrated complete occlusion of the embolized vessel, with a significant reduction of flow into the venous pouch. Small residual shunting vessels feeding from the left PChA and right anterior pericallosal artery remained.

Postoperative Course. The postoperative course was excellent. From a clinical point of view, the baby was doing well, and results of her neurological examination were normal. Congestive liver dysfunction healed dramatically. Follow-up MR imaging also confirmed the effect of the occlusion, showing shrinkage of the vein of Galen ectasia and normal appearance of the vermis. Control angiography studies obtained 12 months after the therapeutic procedure confirmed the disappearance of the AVF (Fig. 1C), demonstrating that the normal veins of the deep system drained into the straight sinus (Fig. 1D). The patient is now 10 years old, developing normally, and does well in elementary school.

Fig. 1. Case 1. Diagnostic ICA angiogram, lateral view, demonstrating a mural-type VGAM fed by the right medial PChA in the arterial phase (A). There is no communication between the galenic pouch and the deep venous draining system in the venous phase (B). Follow-up left ICA angiogram obtained 12 months after the therapeutic procedure demonstrating complete disappearance of the arteriovenous shunts (C). In the venous phase, the left ICV connects to the straight sinus via the shrunken dorsal component of the median prosencephalic vein (D).

**Case 2**

**History and Examination.** This child, the first born to his family, was shown to have ventricular enlargement on prenatal ultrasonography studies performed at 36 weeks of gestation. He was delivered vaginally at 38 weeks and 2 days after a normal pregnancy, his birth weight was 2536 g and head circumference was 35.5 cm. An abnormal pineal cystic lesion was diagnosed on an ultrasonography study obtained at 3 days of age, and the boy was referred for examination at 1 month of age. A plain chest x-ray film revealed no cardiomegaly, and an intracranial bruit was detected on examination. Brain MR imaging and MR angiography demonstrated a dilated vascular structure corresponding to the vein of Galen draining into the falx sinus. The venous pouch enlarged gradually, and congestive liver dysfunction appeared at 4 months.

Operation. At 5 months of age, the child was treated via an endovascular approach. The arteriovenous shunting vessels were located on both sides of the inferior–ventral surface of the dilated venous pouch (Fig. 2A). The venous drainage of the brain was via a persistent primitive accessorial sinus called the falcal sinus in the falx cerebri. There was no evidence of venous reflux from the vein of Galen into the deep veins (Fig. 2B). The biggest shunt, the right medial PChA, was superselectively catheterized and embolized with 0.4 ml of glue. The embolized glue was visualized in the feeding vessel and partially in the ventral component of the ectatic vein (Fig. 2C). Immediate follow-up angiography studies demonstrated complete occlusion of the embolized vessel, with a significant reduction of flow into the vein of Galen and slight improvement in the venous drainage of the healthy brain (Fig. 2D). Small residual shunts feeding from the left PChA remained.

Postoperative Course. The postoperative course was uneventful, and results of neurological examination were normal. Congestive liver dysfunction healed dramatically. Control angiography obtained 6 months after the intervention confirmed the disappearance of the AVF (Fig. 2E), demonstrating that the normal veins of the deep system were draining into the falcal sinus (Fig. 2F). The child has developed almost normally; he is now 9 years old and only has a slight problem in conversation.

**Discussion**

To justify the acceptability of endovascular occlusion, it is generally assumed that the venous aneurysm of a VGAM does not participate in the drainage of normal cerebral structures. Indeed, it is commonly believed that venous structures that normally constitute the deep or Galenic venous system, such as the ICV and the basal vein of Rosenthal, are not connected to the vein of Galen.12 This concept, however, has been questioned in a recent publication. Levrier et al.8 reported on 2 patients with VGAM (a 40-year-old woman and an 11-year-old girl) in whom superselective retrograde transvenous catheterization and MR venography demonstrated that normal double ICVs were draining into the aneurysmal vein of Galen. Gailloud et al.7 reported the confirmation of the communication between the ICV and the vein of Galen by MR venography after endovascular treatment. They emboziled the lesion with glue in 2 sessions (first from
the umbilical route and then from the femoral route) for a VGAM in a newborn baby presenting with severe pulmonary hypertension.

In the case of one of our patients (Case 2), during the control angiography study obtained immediately after embolization, the faint reflux from the galenic pouch into the ICVs and the superior sagittal sinus could be recognized in the diastolic phase. It seemed that these fistulas filled only in 1 direction during the systolic phase or during a pressure arterial injection, whereas during the diastolic phase, and in the absence of a pressure injection, the pattern of drainage temporarily changed. The normal ICV had been present from the beginning but did not show during diagnostic angiographic studies, either because the flow, and therefore the contrast agent, was preferentially directed toward high-flow arteriovenous shunts, or because elevated venous pressure prevented ICV drainage into the aneurysm sac at the time of the diagnostic angiogram. The treatment was subtotal occlusion of the AVF by transarterial glue embolization, which occluded the largest shunt and the ventral aspect of the venous pouch. Even though there was a small persistent flow in both cases, in time both fistulas thrombosed completely. It was demonstrated on the control angiography study obtained after the shrinking of the aneurysmal vein that a single, nondilated ICV draining into the dorsal component of the venous pouch had become apparent. Double ICV could not be confirmed and its presence remains unclear. Both of our reported cases showed the same findings. These results indicate 2 important points. The first is that one must be cautious about completely obliterating the galenic venous pouch in VGAM treatment. In other words, the fact that there are arterIALIZATIONS of normal veins of the brain or, as in our particular cases, no visualization of the normal draining vein, does not mean that the drainage of the fistula can be occluded with impunity. The second point is that VGAMs in babies do not necessarily need to be occluded completely because secondary thrombosis can also occur if the flow is decreased significantly.3

We succeeded in allowing 2 pediatric patients with mural-type VGAM to catch up to their peers in the normal development process by closing an embryonic abnormal cerebral monoshunt with a small amount of glue. The factors for success in this endovascular remodeling technique are as follows: there was no damage in the brain parenchyma at diagnosis, and the systemic manifestation progressed gradually during the observation period; both lesions were mural-type VGAMs for which treatment was postponed until the patients were 5 months (Case 2) and 6 months old (Case 1); a flow-related microcatheter could be introduced into the largest fistulous lesion among the multiple feeding arteries; the liquid embolic material was able to be injected into the biggest vessel fistula as well as the ventral component of the median prosencephalic vein; and other associated feeding arteries closed automatically.

Conclusions

The possibility of such normal deep galenic venous drainage must be considered in the endovascular management of VGAM, because it may imply occurrence of adverse effects when the malformation is occluded on the dorsal component of the galenic pouch with the embolic material. The goal of the endovascular intervention should be to close only the ventral component of the dilated median prosencephalic vein.
References


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