Transumbilical embolization of a congenital dural arteriovenous fistula at the torcular herophili in a neonate

Case report

MASAKI KOMIYAMA, M.D., MISAO NISHIKAWA, M.D., SHOUHEI KITANO, M.D., HIROAKI SAKAMOTO, M.D., NOBUHIRO MIYAGI, M.D., SATOSHI KUSUDA, M.D., and HISAKAZU SUGIMOTO, M.D.

Departments of Neurosurgery, Pediatric Neurosurgery, Neonatology, and Pediatric Cardiology, Osaka City General Hospital, Osaka, Japan

A neonate, in whom a congenital cerebral vascular anomaly had been diagnosed prenatally, exhibited progressive high-output congestive heart failure soon after birth. Cerebral angiography revealed a congenital dural arteriovenous fistula (AVF) with a huge dural lake located at the torcular herophili. In addition to the meningeal blood supply, an unusual pial blood supply from all cerebellar arteries was observed to feed the fistula. The patient was treated by repeated transarterial and transvenous embolization through the umbilical venous route. To the authors’ knowledge, neither the existence of a congenital dural AVF at the torcular herophili presenting with an enormous pial blood supply or the technique of transumbilical venous intervention has been reported in the literature.

KEY WORDS • cerebral angiography • congenital dural arteriovenous fistula • embolization • transumbilical approach • neonate • children

CONGENITAL dural arteriovenous fistulas (AVFs) in children are extremely rare, even rarer than vein of Galen aneurysmal malformations. Congenital dural AVFs may be accompanied by cranial bruits, cardiac failure, dilated scalp veins, macrocephaly, hydrocephalus, delayed neurological development, seizures, or focal neurological deficits—all of which are distinct from symptoms caused by dural AVFs in adults. Congenital dural AVFs and aneurysmal malformations of the vein of Galen are often confused because of their clinical and radiological similarities; however, they are different entities.

For vein of Galen aneurysmal malformations, transarterial embolization, performed through a femoral arterial route, and transvenous embolization, performed either through a femoral or jugular venous route, are commonly used; a transtorcular approach is another option. For congenital dural AVFs, ligation of feeding arteries or surgical resection have been the classic procedures, but recently transarterial embolization has been performed in most cases, occasionally combined with surgical treatment. The transumbilical approach provides unique angiographic routes that are only available in the neonatal period for transarterial or transvenous embolization or both. The purpose of this paper is twofold: 1) to report a rare congenital dural AVF with enormous pial involvement of the cerebellar arteries in a neonate; and 2) to report the performance of transarterial and transvenous embolization through the umbilical vein.

Case Report

The patient’s 27-year-old mother was referred to our hospital after prenatal Doppler sonography indicated a high-flow intracranial vascular anomaly in the fetus at a gestational period of 38 weeks.

Examination. Magnetic resonance imaging performed 3 days before delivery revealed a large signal void area in the posterior portion of the fetal head (Fig. 1A). Because of fetal distress, a cesarean section was performed under application of a spinal anesthetic agent. At a gestational age of 39 weeks, a boy was born with a birth weight of 2801 g. The infant’s Apgar scores were 4 and 9 at 1 and 5 minutes, respectively. His head circumference and height were 34.3 and 48 cm, respectively, which were within the 90th percentile.

Except for cardiac failure and bruits heard over the cervical and occipital regions, the patient was neurologically normal with no anomaly. Because there was no apparent brain damage or hydrocephalus revealed by computerized
tomography scanning performed the day after birth (Fig. 1B), treatment of the patient’s heart failure was warranted. For possible neurointervention, we had attempted to cannulate both of the umbilical arteries and the vein 4 hours after birth. Only the umbilical vein was successfully cannulated using a No. 4 French nutritional tube.

**Diagnostic and Therapeutic Angiography.** On the 4th postnatal day, the first transumbilical venous–transarterial diagnostic and therapeutic angiography was conducted. The No. 4 French nutritional tube in the umbilical vein was replaced by a No. 5 French vascular sheath with a length of 6 cm. This vascular sheath was finally removed on the 17th postnatal day, following the fourth intervention. A No. 5 French balloon-tipped double-lumen catheter (a wedge-pressure catheter; Arrow International, Reading, PA) was navigated from the umbilical vein to the inferior vena cava through the ductus venosus and on to the right atrium, then to the left atrium through the foramen ovale and on to the left ventricle, and, finally, to the ascending aorta (Fig. 1C). Using a long guidewire (0.025 in wide and 260 cm long [Radifocus; Termo, Tokyo, Japan]), the balloon catheter was exchanged for a Tracker-38 catheter with an 18-cm distal flexible port (Target Therapeutics, Fremont, CA), which was introduced into the ascending aorta and on into the brachiocephalic vessels (bilateral common carotid arteries and left vertebral artery). Control angiography was performed using a biplane digital subtraction angiography system (DFP-60A; Toshiba, Tokyo, Japan). The main feeding arteries were the bilateral middle meningeal arteries, bilateral occipital arteries, bilateral tentorial arteries, bilateral superior cerebellar arteries, and bilateral anterior inferior and posterior inferior cerebellar arteries. The occipital and marginal sinuses were patent bilaterally. There was no stenosis or occlusion of the transverse sinuses and sigmoid sinuses or internal jugular veins. Because of the limited dose of contrast material and the poor medical status of the patient, the angioarchitecture of the lesion was not fully understood until three angiographic sessions had been completed (Fig. 1D–F).

---

**Fig. 1.** A: Magnetic resonance T2-weighted image obtained 3 days before delivery while the mother held her breath, revealing a large flow-void area (asterisk) in the posterior portion of the fetal head. The falcine sinus is also shown (arrow). B: Contrast-enhanced computerized tomography scan obtained on postnatal Day 1 demonstrating a huge enhanced structure in the posterior fossa (asterisk) and numerous cerebellar arteries. C: Plain chest–abdominal x-ray film showing the course of the catheter as it passes through the umbilicus, umbilical vein, ductus venosus, inferior vena cava, right atrium, foramen ovale, left atrium, left ventricle, ascending aorta, and on to the right common carotid artery. Note marked cardiomegaly due to high-output failure. Arrow indicates the tip of the vascular sheath placed in the umbilicus. D (early phase) and E (late phase): Angiograms with left internal carotid injection (anteroposterior view) demonstrating the large dural AVF fed by the superior cerebellar arteries (arrowheads) connected through the posterior communicating artery. Note the marked tortuosity in the proximal portion of the internal carotid artery (arrow). F: Angiogram obtained with selective injection (lateral view) to the left middle meningeal artery. A direct fistula to the dural lake at the torcular herophili is clearly shown. G: Plain skull x-ray film (lateral view) showing the deposited platinum coils (total length 590 cm) in the huge dural lake at the torcular herophili as well as in the right occipital artery and the bilateral middle meningeal arteries.
**Embolic Procedures.** We first attempted transumbilical venous–transarterial embolization; however, because of the elongation and coiling of the common carotid and proximal internal carotid arteries as well as their small sizes, embolization was only accomplished in the right occipital artery and bilateral middle meningeal arteries by using platinum coils and in the left anterior inferior cerebellar artery, using polynvinyl alcohol particles (300–500 μ) through a FasTracker 18 microcatheter (Target Therapeutics). The sites for occlusion with coils were proximal to the fistula site, but the infant’s progressive heart failure prompted us to undertake proximal coil placement over three sessions, on the 4th, 10th, and 12th postnatal days. Although the guiding catheter (Tracker 38 catheter) made a loop in the heart to reach the arterial side from the venous side, this loop itself was not a major obstacle for selective catheterization using a microcatheter to reach the targeted vessels and to perform embolization.

To investigate normal cerebral venous flow, a No. 4 French Berenstein catheter was advanced from the umbilical vein into the inferior vena cava through the duc-
tus venosus, on to the right atrium and the superior vena cava, and then, finally, to the right internal jugular vein.

Through this catheter, a FasTracker 18 microcatheter was navigated into the superior sagittal sinus via the right occipital sinus. Selective superior sagittal sinography showed the falciene sinus draining normal venous blood from the cerebral hemispheres to the tentorial sinuses and then to the cavernous and transverse sinuses. Because normal cerebral venous flow returned through the falciene–tentorial sinus route, staged transvenous occlusion of the huge venous pouch at the torcular herophili was warranted. Due to the infant’s heart failure, which progressed even after three sessions of transarterial embolization, we were required to perform transvenous embolization of the huge venous lake through the umbilical venous route on the 17th postnatal day.

**Table 1**

<table>
<thead>
<tr>
<th>Authors &amp; Yr</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gordon, et al., 1977</td>
<td>M</td>
<td>heart failure</td>
<td>ECA ligation</td>
<td>death</td>
</tr>
<tr>
<td>Ross, et al., 1986</td>
<td>M</td>
<td>heart failure</td>
<td>none</td>
<td>death</td>
</tr>
<tr>
<td>Chan &amp; Weeks, 1988</td>
<td>F</td>
<td>heart failure</td>
<td>ligation &amp; resection</td>
<td>alive</td>
</tr>
<tr>
<td>Tessler, et al., 1989</td>
<td>—</td>
<td>heart failure</td>
<td>embolization</td>
<td>death</td>
</tr>
<tr>
<td>Dion, 1993</td>
<td>—</td>
<td>heart failure</td>
<td>embolization</td>
<td>death</td>
</tr>
<tr>
<td>Miller &amp; Albright, 1993</td>
<td>M</td>
<td>heart failure</td>
<td>ligation &amp; resection</td>
<td>death due to tumor</td>
</tr>
<tr>
<td>Lasjaunias, et al., 1996</td>
<td>F</td>
<td>heart failure</td>
<td>—</td>
<td>transient neuro symptom</td>
</tr>
<tr>
<td>M</td>
<td>macrocrania</td>
<td>no embolization</td>
<td>minimal/no symptom</td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>macrocrania</td>
<td>ventricular shunt</td>
<td>severe neuro symptom</td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>seizure</td>
<td>—</td>
<td>death</td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>heart failure</td>
<td>—</td>
<td>minimal/no symptom</td>
<td></td>
</tr>
<tr>
<td>present case</td>
<td>M</td>
<td>heart failure</td>
<td>embolization</td>
<td>no deficits</td>
</tr>
</tbody>
</table>

* ECA = external carotid artery; neuro = neurological; — = not specified.

**Discussion**

**Classification and Angioarchitecture of Congenital Dural AVFs**

Congenital dural AVFs have been reported in the literature in fewer than 50 patients.1,3,6,8,12,15,18,22,27,30,31,35,36,39 Congenital dural AVFs have been hypothetically divided into three subgroups by Lasjaunias, et al.:22 1) dural sinus malformations with arteriovenous shunts; 2) infantile-type dural AVFs; and 3) adult-type dural AVFs. In infantile-type dural AVFs, the dural sinuses are large and patent with no venous lakes. Adult-type dural AVFs are usually located in the cavernous sinuses. Although dural sinus malformations with arteriovenous shunts are observed in all age groups, they are usually found in neonates. Infantile dural AVFs are observed in infants, whereas adult-type dural AVFs are observed in older children.22 Dural sinus malformations with arteriovenous shunts are further divided into two subtypes, one involving the posterior dural sinuses and another involving the jugular bulb. These subtypes have giant dural lakes and slow-flow mural arteriovenous fistulas. Most dural AVFs in neonates are accompanied by cardiac manifestations. The mortality rate in patients with congenital dural AVFs has been reported to be 31 to 38%, whereas that in neonates has been reported to be 67%22,29 Table 1 provides a listing of neonates with congenital dural AVFs at the torcular herophili reported in the literature.4,8,12,18,27,35,39

Congenital dural AVFs at the torcular herophili generally have large meningeal feeding vessels (middle meningeal arteries, occipital arteries, posterior meningeal arteries, and tentorial arteries) and a huge venous lake at the torcular herophili.1,12,18,29,35,39 Pial participation from distal branches of the middle or posterior cerebral arteries,1,12,18,29,35,39 has also been observed, but proved to be less contributory to the AVFs than the menin-
geal feeding vessels. Involvement of all cerebellar arteries, which occurred in our patient, has not been reported. This pial blood supply mimicked the arterioarterial networks (mazes) occurring in choroidal-type vein of Galen aneurysmal malformations. Thrombosis of the dural lake may restrict venous drainage, leading to intraparenchymal hemorrhagic infarction.22

Lasjaunias, et al.,22 postulated that the dural sinus malformation in congenital dural AVFs is attributable to an abnormal fetal development of the sinuses, that is, a persistence of the ballooning of the transverse and/or posterior portion of the superior sagittal sinus, which is a normal phase of sinus development during the 4th to 6th fetal months.32

Treatment of Congenital Dural AVFs

With the advent of interventional neuroradiology, dural AVFs have come to be treated by transarterial embolization instead of by ligation of the feeding vessel or surgical resection. The transfemoral approach has been commonly used. Catheterization of the femoral artery in neonates is challenging and is associated with thromboembolic complication and/or subsequent occlusive changes in the femoral artery.2,7,30 There are limits to the number of repeated interventional procedures that may be undertaken through the same femoral vascular sheath within a few days, because of the difficulty in maintaining femoral arterial flow and the increased possibility of thromboembolic complication. Excessive tortuosity of intracranial and extracranial vessels hampers transarterial embolization of the lesions.13,25 as was the case in our patient. Transarterial glue embolization was an alternative, but because of vascular tortuosity and difficulty in placing glue casts in appropriate locations in the arteriovenous shunts, we did not use this method (and, thus, we did not use a flow-guided microcatheter).

Although using a transvenous approach is technically easier than using the arterial approach, a transfemoral venous approach may still require an arterial catheter for control angiography to demonstrate the angioarchitecture of the dural AVF. Because a huge dural lake connects with other dural sinuses and drains normal cerebral veins, this venous lake should be preserved. Thus, a transvenous approach to occlude the involved sinuses is precluded.22 Only if alternative venous pathways for normal venous return are apparent can transvenous embolization be performed. In our patient, selective sinograms obtained in the superior sagittal sinus showed the falcine–tentorial venous route draining the normal cerebral venous return. After the limited success of transarterial embolization to reduce shunt flow, we performed partial transvenous occlusion of the huge venous lake. Because abrupt, total occlusion of the venous side could have caused catastrophic hemorrhagic complication, we attempted to occlude the venous lake in a graded fashion, anticipating redistribution of venous flow through other channels.25,29 Partial embolization may be useful to improve heart failure in the neonatal period.16 Subsequent embolization can be performed months or years later, following further growth and an improvement in the general condition of the patient.29 Although direct puncture of the venous pouch through the posterior fontanelle and transvenous embolization are theoretically possible, we believe that a transfemoral or transumbilical venous approach is technically easier and safer than using this direct puncture method.

Anatomy of the Umbilical Vessels

The umbilical cord has two thick-walled, round umbilical arteries and one larger thin-walled, oval umbilical vein (Fig. 2).21 The umbilical arteries originate from the internal iliac arteries, run caudal along the sides of the bladder, and turn cephalad along the abdominal wall to the umbilicus. The umbilical arteries constrict rapidly after birth in normal neonates, whereas they remain patent for longer periods in newborns suffering from hypoxia.20 The umbilical vein is located at the 12 o’clock position at the level of the abdominal wall, runs cephalad to the left portal vein, and courses through the ductus venosus, connecting to the inferior vena cava.20,34

Umbilical Approaches for Angiography

Umbilical vein cannulation was first performed by Diamond, et al.,11 in 1946 for exchange transfusion. The first umbilical artery cannulation has been attributed to one undertaken in the late 1950s by Dr. Virginia Apgar, who devised the neonatal resuscitation scoring system.4
Rudolph, et al. reported catheterization through the umbilical vein to reach the heart for hemodynamic and cineangiographic studies in 1961. Umbilical artery or vein catheterization became common in the 1960s. Nonselective transumbilical venous angiography for aortography or cardioangiography and transumbilical arterial angiography for aortography or arteriography, including cerebral vessels, were also first performed in the 1960s. Selective transumbilical arterial angiography using the Seldinger method for thoracoabdominal lesions was first reported in 1977. Selective cerebral angiography for either diagnostic or therapeutic purposes, however, was not performed through the umbilical route until 1997.

**Transumbilical Arterial Approach**

Cannulation of the umbilical artery should be conducted immediately after birth because it is almost impossible after postnatal Day 4. Because the direction of the artery near the umbilicus is caudal, this approach is not convenient for manipulation of catheters for angiography or for intervention. In addition, x-ray exposure to the angiographer cannot be avoided. This approach, however, enables direct access to the aorta and brachiocephalic vessels. Berenstein and colleagues reported that they used this approach for diagnostic and therapeutic angiography focused on vein of Galen aneurysmal malformations. Complications of umbilical artery catheterization include vascular perforation, thromboembolic complications, infection, aortic aneurysm, air embolism, hemorrhage, bladder rupture, and intestinal perforation.

**Transumbilical Venous Approach**

Cannulation of the umbilical vein is easier than that of the artery and may be performed up to at least 7 days after birth. Because the direction of the vein is cephalad, this approach is convenient for diagnostic and interventional procedures. Placement of a catheter in the umbilical vein may be tolerated over a longer period than such a procedure in the umbilical artery. In a transarterial approach, the catheter must pass through the heart, from the right atrium to the left atrium, through the foramen ovale, and on to the left ventricle. There is a risk of inducing arrhythmia during intracardiac catheter manipulation, but this is usually tolerated when a gentle maneuver is used. Both a transjugular venous cerebral approach and a transcardiac–transarterial cerebral approach can be made via the umbilical venous route. To our knowledge, this approach has not been reported in the literature. Complications of umbilical vein catheterization include infection, vessel perforation, thrombosis, liver infarction, cardiac tamponade, and pericardial effusion, cardiac arrest, air embolism, and esophageal varices.

This transumbilical venous–transcardiac catheterization cannot be performed without the cooperation of pediatric cardiologists. We believe that intracardiac catheter manipulation should be performed by pediatric cardiologists who are familiar with transfemoral–transcardiac procedures.

**Conclusions**

We report the case of a neonate with a rare congenital dural AVF of the posterior fossa. This patient was treated by staged transarterial and transvenous embolization. The transumbilical venous approach provides both arterial and venous access to lesions in neonates requiring repeated endovascular interventions.

**References**

Congenital dural AVF treated by transumbilical approach


Manuscript received September 8, 1998. Accepted in final form December 8, 1998.

Address reprint requests to: Masaki Komiyama, M.D., Department of Neurosurgery, Osaka City General Hospital, 2-13-22, Miyakojima-Hondouri, Miyakojima, Osaka 534-0021, Japan. email: komiyama@japan-mail.com.