Neonatal dural arteriovenous fistulas (DAVFs) are rare, but if left untreated will advance to life-threatening neurological and cardiovascular compromise. Endovascular treatment is the preferred treatment modality for DAVFs. The goal of endovascular therapy is to obliterate feeding vasculature and prevent secondary complications. Endovascular access can be difficult to obtain in a neonate. The authors present the case of a full-term, normal birth weight neonate with severe congestive heart failure secondary to a congenital DAVF of the torcular herophili that was successfully treated with transumbilical arterial coil embolization and a liquid embolic agent.

**Keywords** neonatal dural arteriovenous fistula; transumbilical cerebral angiogram; coil embolization; Onyx embolization; vascular disorders; congenital

Endovascular access can be difficult to obtain in neonates. Transfemoral access is traditionally used in neonates weighing more than 2700 g. In low birth weight neonates, the femoral arteries may be too small for a 4-Fr catheter. Even in normal weight neonates, femoral catheterization can lead to thromboembolic and ischemic complications of the leg. In low birth weight neonates with DAVFs, flow in the descending aorta may be compromised, further complicating endovascular access. In patients with birth weights less than 2700 g, obtaining access through other vessels, such as the umbilical arteries, is an alternative. Accessing the umbilical vessels must be obtained within the first week of life before the vessels obliterate.

The materials typically used for embolization in pediatric DAVFs are N-butyl-2 cyanoacrylate (NBCA; ev3/Covidien), Onyx (Covidien Neurovascular, Johnson & Johnson), or detachable coils. Coil embolization complications are observed typically less frequently than with liquid embolic agents because of the possibility of distal agent migration. Coil embolization in combination with a liquid embolic agent with the goal of arterial flow obliteration has been utilized with results indicating relatively lower complication rates. In addition, complete obliteration of the DAVF is not absolutely necessary to significantly reduce the degree of shunting. The combination of the two techniques is plausible and has been published in...
a few cases in the neonatal population, but unfortunately both with poor outcomes.\textsuperscript{4,21}

The low incidence of neonatal DAVFs coupled with poor outcomes despite current best therapy justifies reporting novel strategies for successful treatment. Therefore, we present a case of a full-term, normal birth weight neonate with a congenital DAVF of the torcular herophili that was successfully treated with transumbilical arterial coil and Onyx embolization due to refractory advanced heart failure.

Case Report

History and Examination

The patient was born to a 25-year-old woman at 37 weeks and 3 days of gestation via cesarean section weighing 2800 g. At 20 weeks of gestation, prenatal ultrasonography demonstrated a cystic enlargement extending from the tentorium to calvaria measuring $3.7 \times 2.1 \times 2.0$ cm. A fetal MR image was obtained at 26 weeks of gestation.

At 35 weeks of gestation, the patient had a cardiothoracic area ratio measuring 41%, with increased cardiac ejection velocities and dilated cardiac chambers. The patient was delivered without evidence of macrocephaly, facial venous distension, or carotid bruits.

At birth, the umbilical artery was cannulated to potentially utilize the vessels for future angiography. Ultrasonography demonstrated that the torcular varix measured $5 \times 3 \times 3$ cm with turbulent flow and a fetal arterial waveform within the areas of turbulence. MRI and MR angiography (MRA) performed on the day of birth showed a significant DAVF, bridged by a persistent falx sinus and truncated inferior sagittal sinuses (Fig. 1). The DAVF was fed by a large posterior branch of the right middle meningeal artery (MMA) and multiple external carotid artery (ECA) branches. The patient remained at neurological baseline since birth and without any focal deficits, but was decompensating hemodynamically. The arteriovenous shunting led to worsening right-sided chamber dilation and increases in pulmonary pressures with dependence on prostaglandin E1 to maintain ductus arteriosus patency. He progressed to medically unstable high-output heart failure in the intensive care unit over the next 2 weeks, requiring prostaglandin, furosemide, and milrinone to maintain hemodynamic stability, triggering attempts at endovascular treatment.

Stage I Endovascular Embolization and Postoperative Course

At 16 days old, endovascular treatment was pursued from the preserved umbilical artery access. The patient’s
umbilical artery line was exchanged for a 4-Fr flexible sheath over a 0.018-inch angled tapered microwire, and a 4-Fr diagnostic catheter was used for catheterization of the right internal and external carotid arteries.

After visualizing significant arteriovenous shunting through multiple ECA branches to the distal superior sagittal sinus with a nidal and direct-shunting component, coil and Onyx embolization of the feeding MMA was undertaken. Coil embolization (ev3/Covidien) was first initiated with a series of 8 coils deployed, followed by Onyx embolization of the targeted vessel with Onyx 18 and Onyx 34 (Orbit Galaxy G2 Complex; Johnson & Johnson), allowing minimal reflux. Follow-up imaging showed no appreciable flow through the right middle meningeal vessel, with marked interval decrease in size and arterIALIZATION of the partially thrombosed right peritornical varix. There was a small amount of retrograde arterialized flow from ipsilateral occipital and posterior meningeal branches distally (Fig. 2). The umbilical artery access was maintained with a heparin infusion in anticipation of subsequent embolization. MRA after stage I embolization demonstrated the presence of a large varicose/aneurysmal DAVF of the torcular herophili with a small infarction within the left middle cerebral artery (MCA) territory.

Stage II Endovascular Embolization and Postoperative Course

Two days following the first embolization, the patient’s umbilical artery, which maintained patency from the heparin infusion, was cannulated with a 4-Fr diagnostic catheter. Two large feeders, the right occipital and posterior meningeal arteries, were noted to be contributing to the continued presence of the arteriovenous fistula. A
A series of 0.010-inch X-pedion microwire and Apollo microcatheters were used to access the shunting vessel. Each large vessel was sequentially embolized with Onyx (Fig. 3). Imaging thereafter demonstrated a significant reduction in the dilated torcula, with subsequent obliteration of the MMA feeders.

Postembolization Course

The patient was discharged weighing 4165 g after a 66-day hospitalization. At 2 months’ follow-up, he was able to retain items in his hands for 2–3 seconds. He subsequently recovered, successfully meeting his developmental milestones at 7 and 10 months of life. Follow-up MRI, MRA, and MR venography at 10 months of life indicated new left occipital artery feeder and left ventricular dilation, likely due to chronic steal phenomenon (Fig. 4).

Discussion

We present a case of a successfully treated DAVF of the torcular herophili in a full-term, normal birth weight neonate who had medically intractable cardiac failure using transumbilical cannulation and combined coil and Onyx embolization.

Transfemoral access in normal birth weight infants is reserved as the first option. Varying birth weights have been described to delineate when femoral access may be difficult.9,17 Femoral vessel catheterization can be difficult in neonates of preterm status and of especially low birth weight (< 1500 g), making alternative vascular access points preferable.28 Additionally, transfemoral cannulation of the neonate is associated with stenosis, occlusion, and thrombosis of the iliofemoral vessels more than any other age group. As a result of these complications, future treatments of persistent vascular malformations may become more difficult.19 Transumbilical access within the first week of life for full-term neonates was described in 1970 by Kitterman et al.6 In the neonatal cardiac literature, Linde et al. described successful cannulation of the umbilical artery in an 8-day-old patient, but none thereafter.13 In 1997, Berenstein et al. was the first to utilize the umbilical arteries for endovascular treatment of cerebral arteriovenous malformations.2 Komiyama et al. demonstrated the utility of cannulating the umbilical vessels in a neonate weighing 2801 g.8 This access had been reported in low birth weight, preterm neonates in cerebrovascular malformations typically weighing less than 2800 g.12,17 Our patient weighing 2800 g received umbilical artery cannulation on the first day of life without complication. Adding to the singular case report by Komiyama et al. who treated the normal birth weight neonate, we believe this case revisits the need to consider the umbilical vessels as endovascular access points in all neonates, regardless of birth weight.8 However, the challenges with this technique cannot go unnoticed. Unless the cannulation of the umbilical arteries occurs within the first week of life, patience becomes difficult. With proper umbilical cord care in the immediate postpartum period via the insertion of a standard umbilical arterial line with a heparin drip, the umbilical artery patency can be maintained for extended periods of time while awaiting neurointervention. In particular, adherence to this principal allows neonates who are not born in care centers with proper neonatal critical care services to be transferred to centers with proper endovascular capabilities for treatment without losing the narrow window of umbilical artery patency. An additional utility of the immediate postpartum umbilical arterial line is that it allows for valuable and accurate hemodynamic monitoring in these critically ill patients. We demonstrate a circumstance in which cannulation was performed on the first day of life, but angiography was not performed until 16 days of life. This feature was also unique due to the fact that we maintained patency of the umbilical artery for a significant length of time without complication, such as...

**FIG. 3.** Transumbilical cerebral angiogram of the patient at 18 days old. **A:** Anteroposterior view of the early arterial phase of right common carotid injection after combined coil and Onyx embolization of the right occipital artery (solid arrow) and deep unnamed feeder (open arrow) from the ECA. The previous MMA occlusion (arrowhead) remains obliterated. **B:** Lateral view of the same injection showing the stable MMA occlusion (arrowhead), no filling of the torcula (solid arrow), and obliterated deep feeder (open arrow). Figure is available in color online only.
infection. In neonatal critical care, umbilical vein catheterization may be performed as late as 2 weeks for rapid infusion therapy if the umbilical stump is maintained and cared for, although this length of time has not been observed for endovascular purposes. Additional studies demonstrate the utility of the umbilical artery catheter for follow-up angiography.

Given the successful treatment of our patient, as well as several others presented in additional studies, the transumbilical approach may be a safer option than transfemoral in neonates, regardless of birth weight. Challenges of the umbilical artery approach include vessel tortuosity and the angle of entry requiring the utilization of flexible catheters and wires for the intervention. The risks and benefits of utilizing the transumbilical approach in neonatal endovascular therapy must be weighed against those involved when attempting to cannulate the femoral vessels.

In the pediatric population, the utilization of coiling followed by glue for embolization of DAVFs needs continued investigation. This case report is also one of the very few to demonstrate the successful utilization of combined embolization treatments in a neonatal DAVF. Kincaid et al. reported 3 cases of DAVFs in neonates, all treated with microcoils and NBCA; one died of an acute hemorrhage, one was lost to follow-up, but the third was cognitively normal at 1 year of life. Oshiro et al. utilized the transumbilical approach for glue embolization treatment of a DAVF of the torcular with 4 years of follow up, and the result was a neurologically normal patient. A major concern following glue embolization is the risk of glucosylate deploying into normal vasculature. Berenstein et al. in 1997 and Kincaid et al. demonstrated the possibility of glue emboli dislodging and circulating through the vasculature. Nonetheless, the risk of the intervention must be weighed against the benefits of the intervention. The concern for progressive heart failure was the impetus for treatment of this infant. A systematic framework for treating DAVF has not been published; however, several scoring systems within cerebral arteriovenous vascular anomalies have been described to aid in the decision-making process to intervene in infants younger than 1 month, including the Bicêtre score for vein of Galen malformations. Despite suffering a left MCA infarction after the first coil and glue embolization, our patient has been recovering with physical therapy and occupational therapy throughout the postoperative period. We believe the chronic steal phenomenon leading to left ventricular dilation is a result of flow diversion blood from a high-pressure system into a low-resistance bed, such as the venous system. The ventricular dilation, therefore, is a result of slow atrophy of the left MCA territory cerebrum, creating an ex vacuo effect. Nevertheless, at 10 months of life, our patient has progressed to meet all developmental milestones and his DAVF appropriately treated. We have demonstrated successful treatment of a torcular DAVF after a 2-stage endovascular treatment plan utilizing 1 cannulated umbilical artery with both coil and Onyx embolization.

References

Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
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