A Large Tortuous Umbilical Cord with Superficial Abdominal Wall Varicose Veins in a Newborn

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THE CASE

Term male newborn presented at birth with a varicose umbilical cord and a collateral abdominal circulation.

Prenatal, Birth, and Family Histories

- Born to a 27-year-old gravida 2, para 2 woman
- Nonconsanguinity in the family
- Normal prenatal maternal laboratory screening and fetal survey
- Benign prenatal course
- Delivery at 39 weeks' gestation via vaginal delivery
- Apgar scores 9, 10, and 10 at 1, 5, and 10 minutes, respectively
- Birthweight: 2,810 g (11th percentile); length: 47.5 cm (8th percentile); head circumference: 33.5 cm (15th percentile)

Presentation

After delivery, the infant's umbilical cord appeared to be tortuous and twice the normal width; in addition, large centrifugal varicose veins were noted bulging under the adjacent skin and extending to the anterior abdominal wall (Figs 1–3). The infant appeared otherwise well but was transferred to the NICU for monitoring and further evaluation.

Progression

Physical Examination (Newborn Day).

- General: Active; normal color
- Head and neck: Normocephalic; anterior fontanelle open and flat; neck veins not enlarged
- Lungs: Regular respiratory rate, breath sounds bilaterally equal, no increased work of breathing
- Cardiovascular: Normal S1 and S2, no murmur; femoral pulses present; capillary refill 2 seconds
- Abdomen: Umbilical cord was tortuous and engorged to twice the normal width (no increase in vessel size during crying); centrifugal varicose veins were bulging under the skin and extending to the anterior abdominal wall; soft and nondistended abdomen, with no palpable masses and no hepatosplenomegaly
- Genitourinary: Normal term male genitalia; patent anus
- Skeletal: Equal movement of arms and legs, no visible abnormalities

AUTHOR DISCLOSURE Drs Soares, Freitas, Rodrigues, Lopes, Carvalho, and Pereira have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

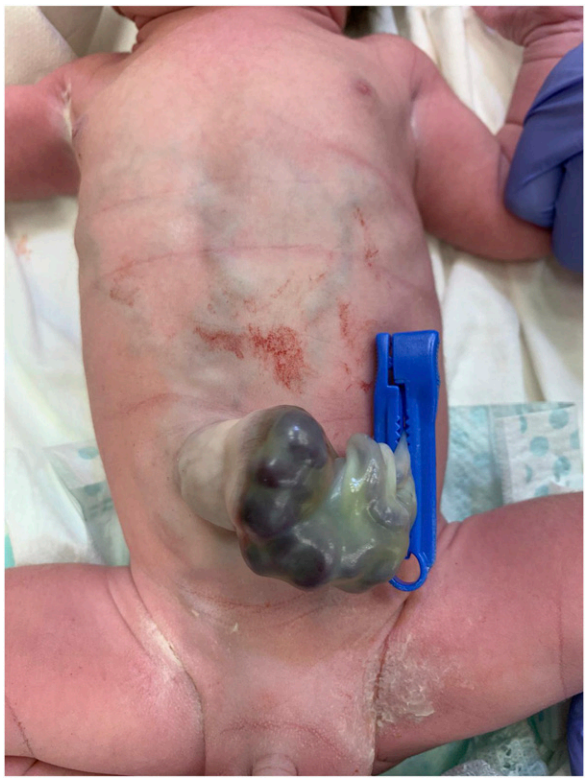

Figure 1. Tortuous and twice the normal width umbilical cord and large centrifugal varicose veins extending to the anterior abdominal wall.

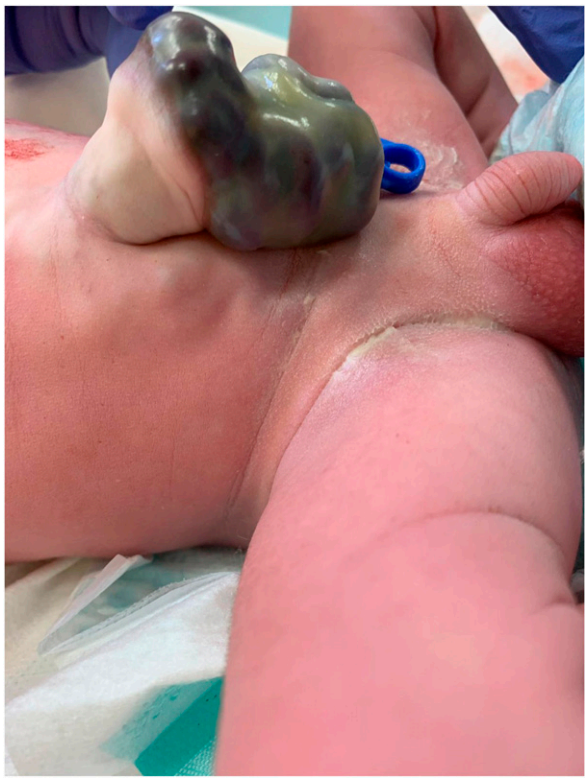

Figure 3. Lateral view of the abnormal umbilical cord.

- Neurologic examination: Appropriate for gestational age with normal tone, strength, and reflexes

Laboratory Studies.

- White blood cell count: $12.7\times10^3/\mu L$ (12.7×10^9/L); hemoglobin: 16.3 g/dL (163 g/L); hematocrit: 46%; platelets: $280\times10^3/\mu L$ (280×10^9/L)
- Blood type: A, Rh positive
- C-reactive protein: Normal
- Blood culture: Negative

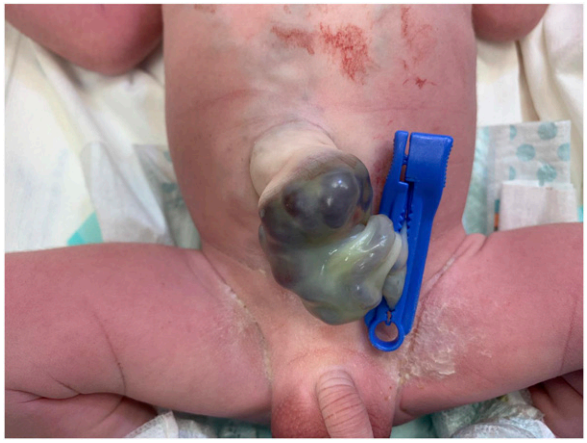

Figure 2. Detailed frontal view.

- 12 hours of age: Alanine transaminase (ALT): 23 U/L (0.38 μkat/L); aspartate transaminase (AST): 79 U/L (1.3 μkat/L); γ-glutamyl transpeptidase: 129 U/L (2.1 μkat/L); alkaline phosphatase: 203 U/L (3.4 μkat/L); total bilirubin: 4.3 mg/dL (73.53 μmol/L); direct bilirubin: 0.52 mg/dL

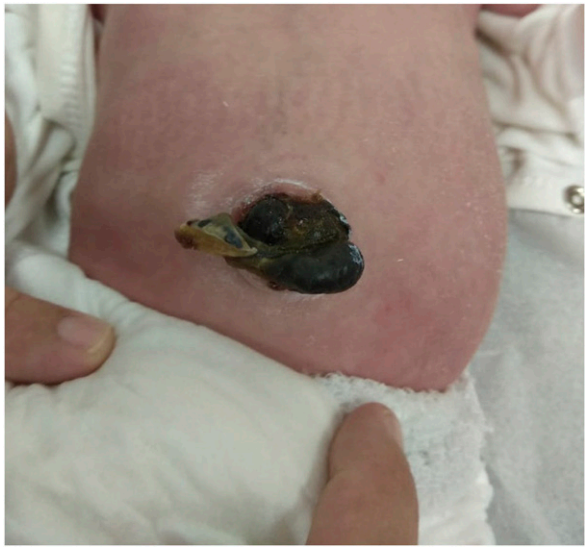

Figure 4. Mummified umbilical stump and little evidence of the collateral circulation.

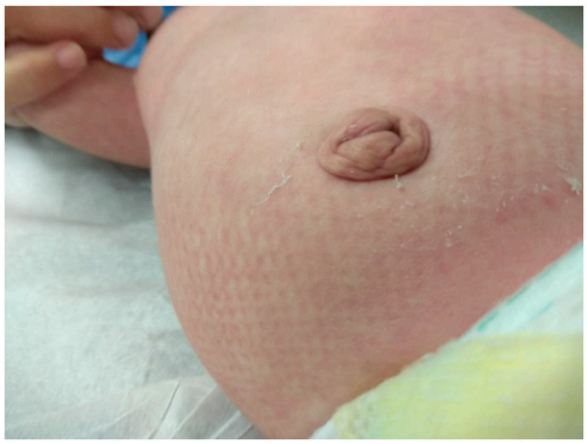

Figure 5. Redundant umbilical tissue.

(8.9 μmol/L); creatine kinase: 735 U/L (12.2 μkat/L); and lactate dehydrogenase (LDH): 723 U/L (12 μkat/L)

- 24 hours of age: AST: 57 U/L (0.95 μkat/L); ALT: 25 U/L (0.42 μkat/L); total bilirubin: 5.65 mg/dL (96.6 μmol/L); direct bilirubin: 0.45 mg/dL (7.7 μmol/L); creatine kinase: 501 U/L (8.3 μkat/L); LDH: 655 U/L (10.9 μkat/L)

Differential Diagnosis

- Caput medusae
- Inferior vena cava obstruction
- Budd-Chiari syndrome (ie, occlusion of the hepatic veins), either primary or secondary
- Portal hypertension

Imaging Studies

- Echocardiography: No structural heart disease, normal function
- Abdominal ultrasonography (Doppler): Anterior abdominal collateral circulation; patent and normally inserted inferior vena cava; normal abdominal organs
- Computed tomography angiography: Anterior abdominal collateral circulation; patent and normally located inferior vena cava, portal vein and suprahepatic veins; normal abdominal organs

Actual Diagnosis

- Caput medusae

MANAGEMENT

After a multidisciplinary discussion with neonatology, radiology, and pediatric vascular surgery, the team decided to take an expectant approach. During the first days after birth, the umbilical cord volume and collateral abdominal circulation gradually decreased. The newborn was discharged from the hospital on the 7th day after birth, with a mummified umbilical stump and little evidence of the collateral circulation (Fig 4).

The umbilical stump fell off on the 20th day of age. At 1 month of age, the infant had redundant umbilical tissue (Fig 5) and small areas in the upper abdominal quadrants showing the collateral circulation. At 4 months of age, there was no evidence of a collateral abdominal circulation.

What the Experts Say

Caput medusae in a newborn is a rare condition with only a few cases reported in the literature. (1)(2) This Latin term translates into "the head of Medusa" and compares the snakes emanating from the head of the Greek monster Medusa with the vessels of the umbilical cord and surrounding abdominal wall. White et al theorized that the caput medusae resulted from the persistence of the right umbilical vein. (1)

During the development of the normal fetal circulation, there are initially 2 paired veins in the embryo: 1) the gut vein and its appendages, known as the omphalomesenteric veins, and 2) the veins from the placenta that form the umbilical veins. In the 4-week embryo, the paired umbilical veins run separately on both sides and drain into the sinus venosus. As the liver increases in size anteriorly, the umbilical veins connect with the omphalomesenteric veins to form large venous lakes in the liver. Some branches of the omphalomesenteric veins anastomose around the gut and the venous liver lakes form the ductus venosus in an opposite location from the entry of the left umbilical vein. Later (at the 7-week embryo stage), several anastomoses of the 2 omphalomesenteric veins occur, leading to all of the gut blood draining to the liver through the portal vein. Part of the omphalomesenteric veins is believed to form the hepatic veins and the hepatic part of the inferior vena cava. As this process continues, the right umbilical vein loses its connections to the superior vena cava and to the liver and is reabsorbed, while the left umbilical vein enlarges to form the mature umbilical vein.

Arrest in this final phase of right umbilical vein occlusion is responsible for the existence of an alternative umbilical circulation, maintaining blood flow from the placenta through the superficial abdominal veins and resulting in the caput medusae. This persistence is not associated with other congenital malformations. After delivery, the umbilical cord is clamped, blood flow from the placenta ceases, and the collateral circulation progressively disappears.

Other causes should be considered in the differential diagnosis of a newborn with an abdominal collateral circulation, including pathologies that require urgent management. These findings can be observed in an infant with obstruction of the inferior vena cava or if there is obstruction

of the hepatic veins (known as Budd-Chiari syndrome). A primary cause of Budd-Chiari syndrome is thrombosis of the hepatic veins and secondary causes are compression or invasion of the hepatic veins and/or inferior vena cava. (3) These other causes can be diagnosed on abdominal ultrasonography with Doppler or computed tomography angiography. Evaluation by cardiology is also essential for exclusion of a cardiac pathology. (3)

SUMMARY

- Caput medusae is a rare neonatal finding.
- Primary or secondary Budd-Chiari syndrome should be excluded in the neonatal period.
- In contrast to adults with caput medusae from portal hypertension, this collateral abdominal circulation can be a benign variant.
- If cardiac or venous malformations are ruled out, an expectant approach is indicated because the collateral veins will gradually involute in the first weeks after birth without sequelae.

American Board of Pediatrics Neonatal-Perinatal Content Specification

- Recognize the diagnostic implications of single vs. multiple anomalies.

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ANSWER KEY FOR NOVEMBER 2020 NEOREVIEWS

Congenital Neuroblastoma: 1. C; 2. B; 3. A; 4. D; 5. E.

Intravascular Hemolysis and Complications During Extracorporeal Membrane Oxygenation: 1. C; 2. D; 3. D; 4. E; 5. B.

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