Interventional Radiology:

Transarterial Embolization of a Hepatic Arteriovenous Malformation in an Infant Using Onyx: A Case Report and Review of the Differential Diagnosis Imaging Findings

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Transarterial Embolization of a Hepatic Arteriovenous Malformation in an Infant Using Onyx: A Case Report and Review of the Differential Diagnosis Imaging Findings

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ABSTRACT

Hepatic arteriovenous malformations are rare congenital lesions associated with significant morbidity and mortality, most commonly from high output cardiac failure. Efficient diagnosis and treatment demands an interdisciplinary approach, and the interventional radiologist plays a pivotal role in both. Imaging is important for diagnostic accuracy and treatment planning, and transcatheter embolization has become an established primary therapy. We report the clinical and imaging findings of a rare hepatic arteriovenous malformation in an infant presenting with high-output cardiac failure and pulmonary artery hypertension that was successfully treated by transarterial embolization using Onyx.

CASE REPORT

CASE REPORT

A term male infant was delivered by a 20-year-old woman following an uncomplicated pregnancy. Shortly after delivery, the infant developed tachypnea, pallor, and poor muscular tone. A harsh systolic murmur was detected on physical exam and oxygen saturation was 74%. The patient required emergent intubation.

Imaging findings:

Profound cardiomegaly and trace bilateral pleural effusions were seen by chest radiography (Fig. 1). Echocardiogram revealed severe right heart dysfunction with atrial septal bowing, marked right atrial enlargement, and paradoxical flow across a patent foramen ovale (Fig. 2). A patent ductus arteriosus with right to left flow and 10 mmHg gradient indicative of pulmonary arterial hypertension was also identified (Fig. 3). Right ventricular function was moderately decreased, but left ventricular function was preserved. A vascular lesion was incidentally found in the liver.

Subsequent duplex Doppler ultrasound (US) of the liver showed no mass, but multiple dilated vascular channels were identified in the right lobe together with marked enlargement of the hepatic artery and veins. Resistive index (RI) in the hepatic artery was decreased indicating increased diastolic flow (Fig. 4). The hepatic veins demonstrated turbulent flow and the spectral Doppler waveform of the inferior vena cava (IVC) was arterialized (Fig. 5).

Magnetic resonance imaging (MRI) of the liver demonstrated a corresponding confluent collection of abnormal blood vessels in the right hepatic lobe measuring 4.6 cm x 4.2 cm x 3.0 cm, but no associated soft tissue mass to suggest a neoplasm. Intralesional flow voids were present within the abnormal vessels in keeping with high flow velocity (Fig. 6).

Management:

The clinical features and imaging findings supported the diagnosis of a hepatic arteriovenous malformation (AVM).

The patient was initially treated medically with furosemide for congestive heart failure and prostaglandin E1 to maintain ductal patency, but cardiac function did not improve. Surgical resection was deemed too risky given the high risk of perioperative hemorrhage and thus mortality. Our interventional radiology group was consulted for additional treatment options.

On day 7 of life a transarterial embolization of the hepatic AVM was performed (Fig. 7). Right transfemoral arterial access was obtained with US guidance using micropuncture technique and a 4 French vascular sheath was placed. The celiac artery was catheterized using a 4 French reverse curve catheter and a coaxial microcatheter was used to selectively catheterize the hepatic arteries. Angiography demonstrated marked hypertrophy of the celiac artery, which fed a high flow vascular malformation in the right hepatic lobe with early draining veins and regions of arterioportal shunting (Fig. 8). Multiple feeding branches derived from the hepatic arteries were identified and embolized using a combination of Onyx 18 and 34 (Fig. 9). Greater than 75% of the AVM was successfully treated, with only minimal residual enhancement of the central nidus on the completion arteriogram (Fig. 10 & 11). No procedural complications were encountered.

Follow-up:

The patient's clinical status improved such that on post-procedure day 2 he was extubated and required less furosemide. The clinical manifestations of right heart overload subsided, cardiomegaly improved by chest radiography (Fig. 12), and there was normalization of the right ventricular function by echocardiography. On day 19 of life he was discharged home on furosemide. At 2 months of life the furosemide was discontinued as the patient was completely asymptomatic. The patient continues to be closely followed as an outpatient, but has a normal physical exam and is meeting normal developmental milestones by 5 months of age.

DISCUSSION

Vascular anomalies can be broadly placed into one of two categories: those of neoplastic origin and those that represent congenital malformations [1]. Vascular anomalies of a neoplastic origin exhibit increased cell proliferation constituting a soft tissue mass, whereas vascular malformations do not. Rather, they solely consist of dysplastic blood vessels that grow in proportion with the child as they mature [2, 3]. Furthermore, vascular malformations can be subdivided into slow-flow (e.g. venous malformation) and high-flow (e.g. arteriovenous malformation) lesions based upon the presence or absence of an arterial component [4, 5]. Accurate diagnosis of vascular malformations is important as specific treatments may vary [2].

Etiology and Demographics

An isolated hepatic arteriovenous malformation as seen in this case is a rarely encountered congenital vascular anomaly mostly limited to case reports [6]. There is no known sex predilection and the etiology and pathogenesis of AVMs are not well understood [7, 8]. AVMs are commonly seen in

hereditary hemorrhagic telangiectasia (HHT) patients, however, a neonatal presentation of an HHT-associated hepatic AVM is also limited to case reports [9]. HHT, also known as Osler-Weber-Rendu syndrome, is an autosomal dominant syndrome associated with angiodysplastic lesion formation in the brain, lungs, liver, skin, and mucous membranes. The etiology is due to mutations in the ENG and ACVRL1 genes. HHT occurs at an incidence of approximately 1:10,000 births, although in some regions of Europe and the Caribbean the incidence is higher [10].

Clinical and Imaging Findings

Arteriovenous malformations are present at birth and grow in proportion with the child. Although congenital, AVMs may initially be clinically silent and present later in life including adulthood. Complications include high-output cardiac failure, hemorrhage, embolism, pain, or the consumptive coagulopathy known as Kasabach-Merritt syndrome [11, 12]. Stigmata of HHT include telangiectasias of the skin and mucous membranes and AVMs of the liver, pulmonary or central nervous system. Symptoms of HHT are broad and attributable to the multitude of angiodysplastic lesions; clinical manifestations include epistaxis, GI bleeding, hypoxia, stroke, and brain abscesses among others [13].

On cross sectional imaging, hepatic AVMs appear as a tangle of dysplastic arteries and veins with imaging characteristics of high blood flow such as rapid enhancement on postcontrast imaging, decreased RI by US, and flow voids on MR [3, 5]. An important diagnostic feature is lack of an associated soft tissue mass, which is present in vascular malformations of a neoplastic origin such as the infantile hepatic hemangioma [2, 3]. Another distinguishing feature of AVMs is the presence of a nidus: the region of abnormal arteriovenous (AV) communication at the AVM core that bypasses the normal tissue capillary bed [4]. It is the nidus that must be occluded during transcatheter embolization to ensure optimal treatment [14].

By grayscale US, hepatic AVMs appear as dilated vascular channels in the parenchyma accompanied by enlarged hepatic arteries and veins. Arterial spectral Doppler interrogation will frequently show high systolic and diastolic flow with resultant low RI. Venous spectral Doppler analysis may show arterialization of venous waveforms due to intralesional AV shunting [4].

Catheter angiography of AVMs will reveal enlarged arteries feeding a nidus at the lesion core with dilated draining veins that rapidly appear. A parenchymal vascular blush is not seen, which typifies a neoplastic process [15].

Contrast enhanced computed tomography (CT) and MRI will demonstrate confluent abnormal blood vessels with brisk enhancement. Regions of AV shunting are responsible for transient hepatic attenuation/intensity differences (THAD/THID) that may be seen on post-contrast CT and MR. Spin echo MR sequences will typically demonstrate vascular flow voids secondary to the high flow nature of these lesions [3, 4].

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Treatment and Prognosis

The prognosis of an isolated hepatic AVM presenting in an infant is poor. Morbidity is high and mortality rates often exceed 50%, most commonly from high-output cardiac failure [11, 16]. Medical therapy is directed at controlling associated conditions such as high output cardiac failure, pulmonary hypertension, and consumptive coagulopathy. Therapies such as radiation, surgery, or transcatheter embolization are targeted directly at the AVM. Transcatheter embolization is performed with the intention of occluding the AVM nidus and can be done via transarterial or transvenous (retrograde) approaches [4, 14]. Surgical management options include ligation of feeding vessels, resection, and liver transplantation. Transcatheter embolization followed by surgical resection is often the mainstay of treatment, although a lack of randomized controlled trials makes the formulation of strict treatment guidelines difficult [11, 17, 18]. Due to the complexity of vascular malformations, it is logical that best management results from an interdisciplinary approach [19].

To the best of our knowledge this case represents the youngest patient reported in the literature with a hepatic AVM successfully treated by transarterial embolization using Onyx (Covidien, Plymouth, MN) [20]. The Food and Drug Administration approved Onyx for treating intracranial AVMs, but it has useful off-label uses [21]. Onyx is a permanent liquid embolic agent available in difference viscosities (i.e. 18, 34, and 500) that is composed of ethylene-vinyl alcohol copolymer (EVOH) with microionized tantalum powder to provide radiopacity. Once Onyx contacts blood, it precipitates into a non-adhesive spongy-like solid that induces endothelial inflammation thereby promoting thrombosis. In order to prevent early precipitation, Onyx must first be dissolved in a solvent of dimethyl sulfoxide (DMSO), which is instilled in the delivery catheter prior to Onyx administration. Unlike tissue glue, Onyx has less risk of adherence to the delivery catheter tip [21]. Onyx was believed to be best choice of embolic agents in this particular case as its viscous nature allowed for precise delivery. This was of major importance due to the high risk of nontarget embolization to the lungs and brain in the setting of known pulmonary arterial hypertension and patent foramen ovale with reversed (paradoxical) flow.

Differential Diagnosis

Unfortunately, a hepatic mass in a child is frequently from metastatic disease. However, age at presentation is important as multiple hepatic masses in a child less than 6 months of age likely represent hemangiomatosis whereas in children older than 6 months of age neuroblastoma metastases are more likely [22]. Primary malignant hepatic tumors are also more common than primary benign tumors in children [23, 24]. When a liver mass is discovered in a neonate, the possible etiologies include hepatic vascular anomalies (infantile hemangioma and very rarely arteriovenous malformation), metastatic neuroblastoma, hepatoblastoma, and mesenchymal hamartoma.

An infantile hepatic hemangioma (IHH) is the most common benign hepatic mass in a newborn [23]. Infantile hepatic hemangiomas are small or occult at birth and tend to involute during childhood, whereas hepatic AVMs are present at birth and grow in proportion with the child [5]. An IHH is a

true neoplasm and appears as a soft tissue mass on imaging. Conversely, AVMs are solely comprised of dysplastic blood vessels without a demonstrable soft tissue mass [5]. By grayscale US, IHHs appear as a hypoechoic mass relative to normal liver and may contain punctate echogenic foci with acoustic shadowing in keeping with calcification. Arteriovenous malformations appear as confluent dysplastic blood vessels without associated soft tissue mass. Doppler US findings are similar in both lesions with low RIs (increased diastolic flow) and arterialized venous waveforms from AV shunting [23]. By noncontrast CT, IHH will appear as a hypoattenuating soft tissue mass often with calcifications. By MR an IHH appears as a T1 hypointense, T2 hyperintense mass and may have foci of increased T1 as a result of intralesional hemorrhage. Enhanced CT and MR images show distinctive early peripheral nodular enhancement with progressive centripetal fill-in on delayed images [23]. In comparison, AVMs appear as a tangle of abnormal blood vessels on CT and MR with rapid arterial enhancement and early draining veins on postcontrast imaging [5, 23].

Neuroblastoma (NB) is the third most common pediatric malignancy, and NB metastases are the most frequently encountered malignant hepatic mass in the neonate [22, 24]. If suspected clinically, elevated urine catecholamines support the diagnosis and are seen in the majority of cases. Serum AFP levels may also be elevated although this is seen less frequently. Affected patients may present clinically with hepatomegaly thereby prompting imaging [22]. By grayscale US, NB metastases appear as hypo to isoechoic masses relative to normal liver, but may be occult if infiltrative disease is present. By CT, NB metastases typically appear as hypoattenuating masses and are T1 hypo- and T2 hyperintense by MRI. Post contrast CT and MR imaging appearance is variable, but neuroblastoma metastases tend to hypoenhance relative to adjacent liver in a heterogeneous fashion [22].

Hepatoblastoma (HB) is the most common primary hepatic malignancy in children [24]. Most patients with HB will have an elevated serum alpha-fetoprotein (AFP), which can aid in the diagnosis. By grayscale US, HB is usually hyperechoic to normal liver but may contain hypoechoic septa. Internal hypervascularity can be seen by Doppler interrogation. On cross sectional imaging, HB appears as a distinct soft tissue mass (in contradistinction to AVMs) that may contain fibrotic septa in a "spoke-wheel" pattern [24]. By CT, HB appears as a hypoattenuating soft tissue mass that frequently has calcifications. By MRI, HB appears as a T1 hypointense, T2 hyperintense mass that may have T1 hyperintense foci of hemorrhage. Heterogeneous enhancement is also typical by both CT and MRI. [24].

A mesenchymal hamartoma is the second most common benign pediatric hepatic mass [23]. It has a distinctly different imaging appearance to that of a hepatic AVM, in that it is a relatively avascular lesion comprised of numerous cysts and solid stromal components. [23]. Grayscale US will demonstrate multiple simple cysts and/or cysts complicated by septa, internal debris, or solid mural components. Doppler interrogation may show internal vascularity within the septa and solid components. CT will show a mass comprised of

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mostly fluid attenuation cysts. Solid components and septa will appear hypoattenuating relative to normal liver parenchyma. On MR the cystic components are T2 hyperintense and show varying degrees of T1 signal intensity that range from hypointense if simple fluid to hyperintense if the fluid is proteinaceous. The solid stroma and septa demonstrate hypointense signal on both T1 and T2 weighted imaging. Stromal components will enhance following contrast administration [23].

TEACHING POINT

Hepatic arteriovenous malformations are rare congenital vascular lesions with significant morbidity and mortality. Catheter directed embolization is an effective minimally invasive means of therapy.

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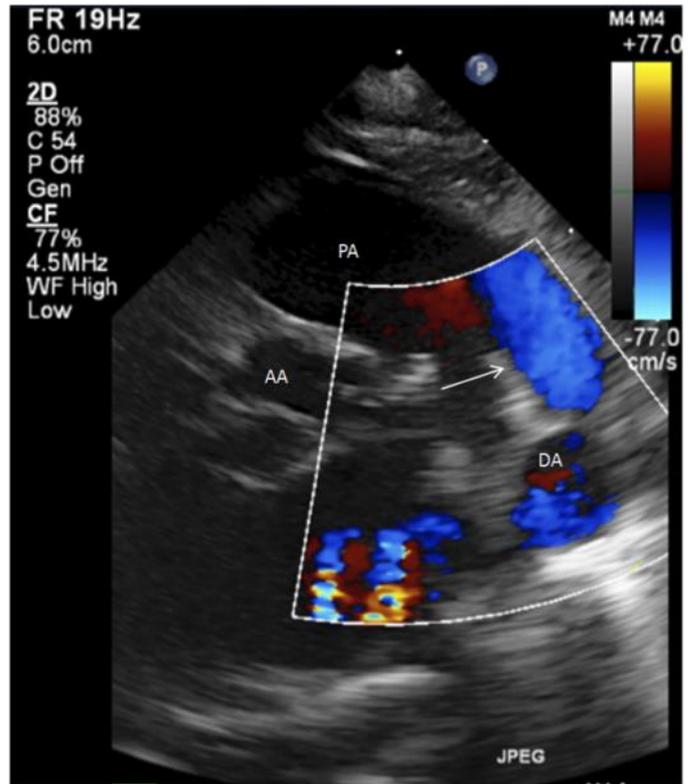
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FIGURES

Figure 1: Conventional radiography.
Male newborn with hepatic AVM.
FINDINGS: Marked cardiomegaly and trace pleural effusions (arrows).
TECHNIQUE: Portable AP chest radiograph. Shimadzu MobileDaRt Evolution.
61 kVp. 1 mAs. 40 inches from patient.

Figure 3: Echocardiography with color Doppler.
Male newborn with hepatic AVM.
FINDINGS: Enlarged pulmonary artery (PA) Boston z-score 2.3 and patent ductus arteriosus with reversed flow (arrow). AA - ascending aorta. DA - descending aorta.
TECHNIQUE: High parasternal view. Philips iE33 ultrasound unit. S12-4 MHz sector array transducer.

Figure 2 (left): Echocardiography.
Male newborn with hepatic AVM.
FINDINGS: Marked right atrial (RA) and right ventricular (RV) enlargement with patent foramen ovale (arrow). LV - left ventricle. LA - left atrium.
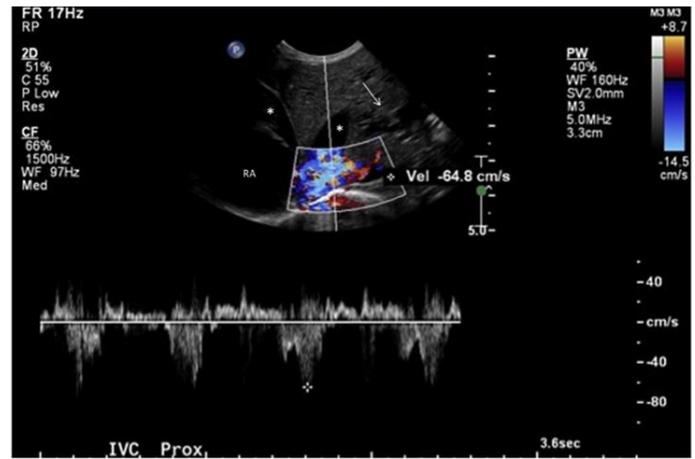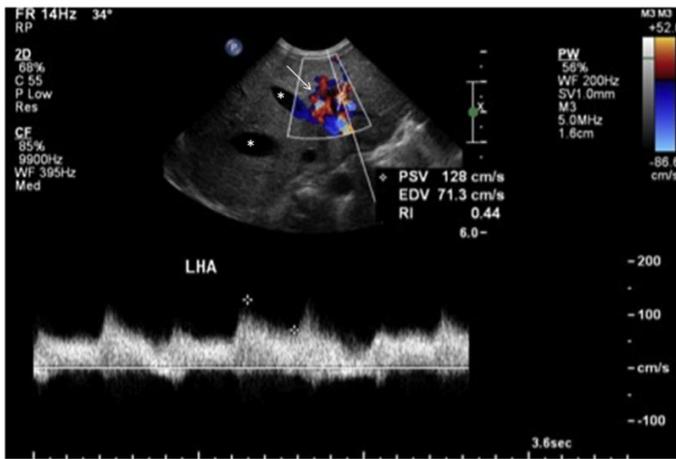TECHNIQUE: Four-chamber view. Philips iE33 ultrasound unit. S8-3 MHz sector array transducer.

Figure 4: Grayscale and Doppler ultrasound of the liver. Male neonate with hepatic AVM.

FINDINGS: Tangle of dysplastic blood vessels (arrow) and dilated (up to 9 mm caliber) hepatic veins (*) with corresponding Doppler flow. Spectral Doppler of left hepatic artery shows high systolic and diastolic flow with low resistive index indicative of intralesional arteriovenous shunting.

TECHNIQUE: Axial view. Philips iU22 ultrasound unit. C8-5 MHz curved array transducer.

Figure 5: Grayscale and Doppler ultrasound of the liver. Male neonate with hepatic AVM.

FINDINGS: Marked right atrial (RA) enlargement, dilated (up to 9 mm caliber) hepatic veins (*) and dilated vascular channels (arrow) in the liver parenchyma. Spectral Doppler shows arterial pulsations in the IVC from arteriovenous shunting.

TECHNIQUE: Sagittal view. Philips iU22 ultrasound unit. C8-5 MHz curved array transducer.
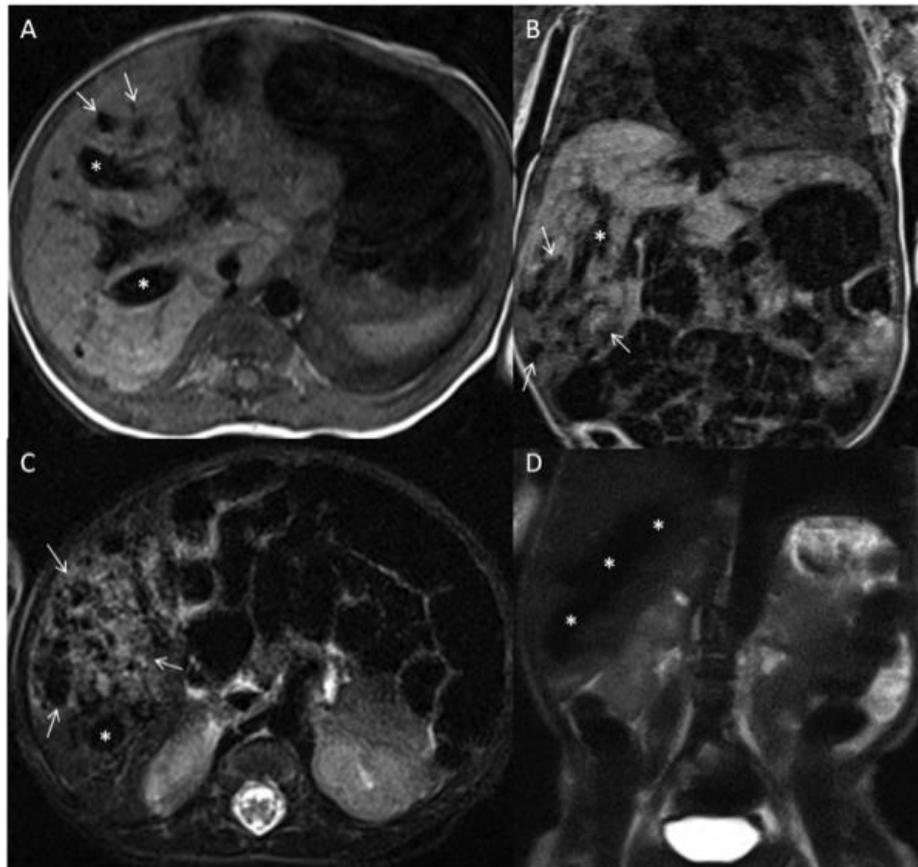
Figure 6: Magnetic Resonance Imaging of the liver. Male newborn with hepatic AVM.

FINDINGS: Enlarged (up to 9 mm caliber) hepatic veins (*) and dilated serpiginous vascular channels with corresponding flow voids (arrows) in the right hepatic lobe. Perilesional increased T2 signal (image C) is consistent with mild congestive edema.

TECHNIQUE: Siemens MAGNETOM Avanto 1.5 T. Noncontrast sequences. (A) axial T1, TR: 477 TE: 15, (B) coronal T1, TR: 460 TE: 9.9, (C) axial T2 fat suppressed, TR: 3470 TE: 140 (D) Coronal T2 fat suppressed, TR: 1690 TE: 98.

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Figure 7 (left): Intraprocedural photograph.
Male newborn with hepatic AVM.
FINDINGS: Anesthesiologist intubates patient following induction of general anesthesia.
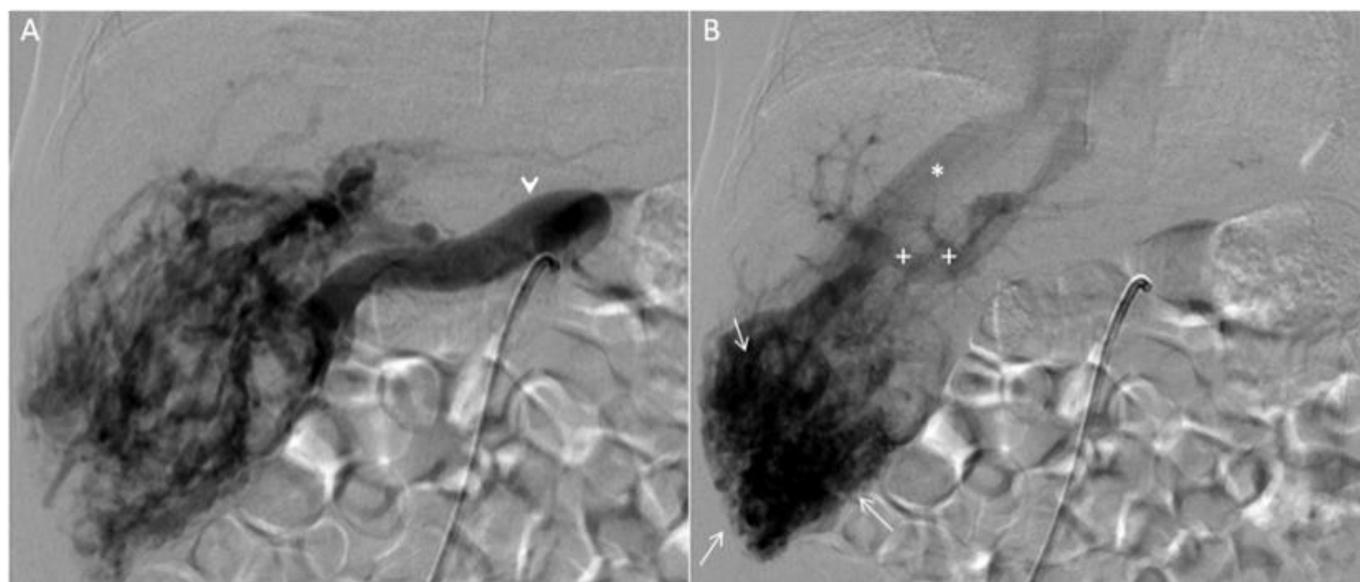TECHNIQUE: iPhone 5 camera, f/2.4, 1/20 sec., ISO-64.

Figure 8: Digital subtraction angiography.
Male newborn with hepatic AVM.
FINDINGS: Hypertrophic (10 mm caliber) celiac (arrowhead) and hepatic artery (7 mm caliber) supplying a vascular lesion with central nidus (arrows) and an enlarged (9 mm caliber) early draining hepatic vein (*) consistent with AVM. Opacification of the portal venous system (+) signifies arterioportal shunting.
TECHNIQUE: Arteriogram of the celiac artery in arterial (A) and parenchymal (B) phases obtained prior to transcatheter embolization. Siemens AXIOM Artis C-arm. Hand injection 5 cc Omnipaque 350. kVp 64, mAs 10.

Figure 9: Fluoroscopic spot images.
Male newborn with hepatic AVM.
FINDINGS: Catheter directed embolization of feeding hepatic arterial branches supplying an AVM using Onyx (arrows) as the embolic agent.
TECHNIQUE: Fluoroscopic spot images obtained early (A) and later (B) in the procedure. Siemens AXIOM Artis C-arm. kVp 66, mAs 13.
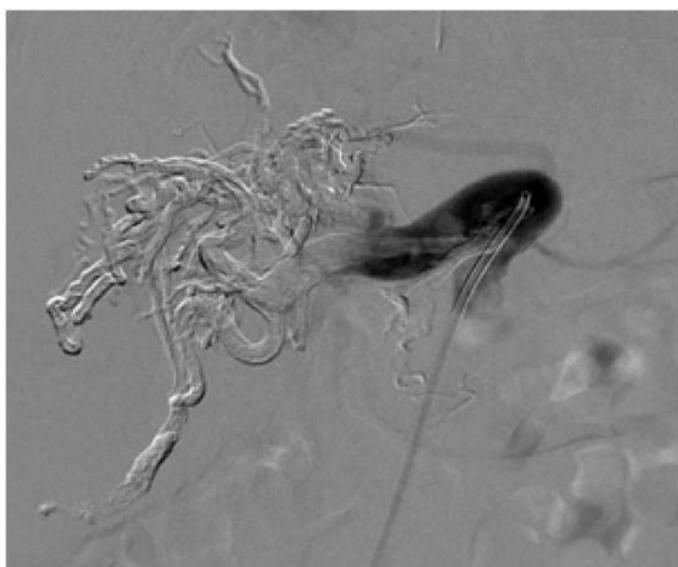

Figure 10: Digital subtraction angiography.
Male newborn with hepatic AVM.
FINDINGS: Near total occlusion of hepatic AVM following catheter directed embolization with Onyx.
TECHNIQUE: Celiac arteriogram. Siemens AXIOM Artis C-arm. Hand injection 5 cc Omnipaque 350. kVp 68, mAs 6.
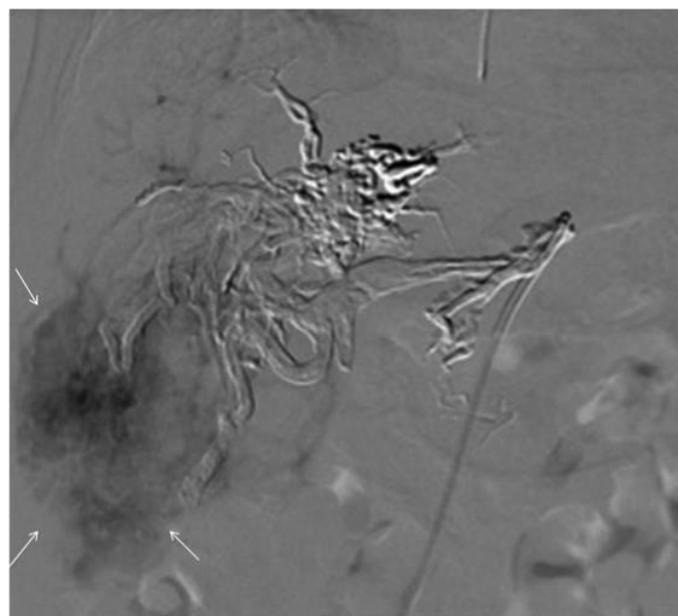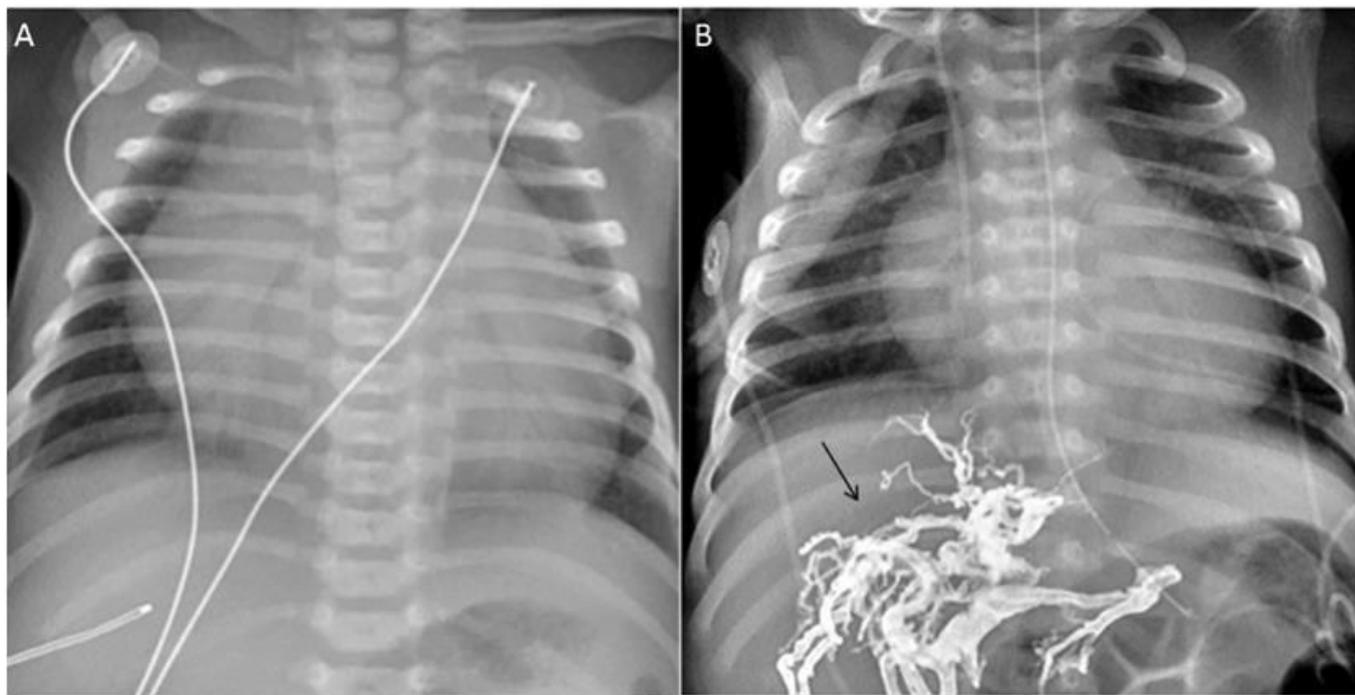

Figure 11: Digital subtraction arteriography.
Male newborn with hepatic AVM.
FINDINGS: Minimal residual filling of the AVM nidus (arrows) following catheter directed embolization with Onyx.
TECHNIQUE: Celiac arteriogram. Siemens AXIOM Artis C-arm. Hand injection 5 cc Omnipaque 350. kVp 68, mAs 6.

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Figure 12: Conventional radiography.

Male newborn with hepatic AVM.

FINDINGS: Presenting cardiomegaly (left) improved following transarterial embolization of hepatic AVM (right). Note radiopaque Onyx embolic material in the hepatic AVM (arrow).

TECHNIQUE: Portable AP chest radiographs before (A) and after (B) transarterial embolization.

Shimadzu MobileDaRt Evolution. 61 kVp. 1 mAs. 40 inches from patient.

Etiology	Unknown.
Incidence	Isolated hepatic AVMs are very rare, limited to case reports; Hereditary Hemorrhagic Telangiectasia (HHT) related hepatic AVMs are more common.
Gender Ratio	No sex predilection.
Age Predilection	Congenital lesion.
Risk Factors	HHT; (neonate presentation of HHT is rare).
Treatment	Medical management; catheter directed embolization, surgical resection, ligation, or liver transplantation; radiotherapy. Definitive treatment with embolization followed by resection.
Prognosis	Poor; approximately 50% mortality for non HHT-associated AVM.
Findings on imaging	High-flow vascular lesion without intervening soft tissue mass comprised of dilated arteries feeding a nidus and prominent early draining veins.

Table 1: Summary table for hepatic arteriovenous malformation (AVM)

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Entity	US	CT	MRI	Enhancement
Infantile Hepatic Hemangioma	• **Grayscale:** Hypoechoic soft tissue mass. May have echogenic, shadowing foci from calcifications. • **Doppler:** High systolic and diastolic flow resulting in low resistive index (RI). Arterialization of venous waveform.	• Hypoattenuating soft tissue mass. • Calcifications are common.	• T1 hypointense, T2 hyperintense soft tissue mass. • Intralesional hemorrhage may appear as T1 hyperintense foci. • May have flow voids on spin echo sequences.	• Distinctive early peripheral nodular enhancement with progressive centripetal fill-in on delayed images.
Metastatic Neuroblastoma	• **Grayscale:** Hepatomegaly. Hypoechoic to isoechoic liver lesions. Infiltrative liver metastasis may be difficult to detect by US. • **Doppler:** May have internal vascularity.	• Hepatomegaly. • Multiple hypoattenuating liver lesions.	• Hepatomegaly. • Multiple T1 hypointense, T2 hyperintense lesions.	• Variable; tends to hypoenhance relative to adjacent normal liver.
Hepatoblastoma	• **Grayscale:** Hyperechoic soft tissue mass with hypoechoic septa. May have echogenic, shadowing foci from calcifications. • **Doppler:** May show internal vascularity.	• Heterogeneous, predominately hypoattenuating mass. • Fibrous septa may create "spoke wheel appearance". • Calcifications are common.	• T1 hypointense, T2 hyperintense soft tissue mass. • Intralesional hemorrhage may appear as T1 hyperintense foci.	• Heterogeneous enhancement pattern.
Mesenchymal Hamartoma	• **Grayscale:** Multiple anechoic cysts with hyperechoic solid mural components and intervening septations. • **Doppler:** Solid components and septa may show internal vascularity.	• Multiple cysts of differing sizes that may contain septations. • Calcifications uncommon.	• T1 variable. T2 hyperintense cysts and T2 hypointense septations.	• Enhancing septa and stromal components.
Hepatic Arteriovenous Malformation (AVM)	• **Grayscale:** Confluent dysplastic blood vessels without associated soft tissue mass. • **Doppler:** High systolic and diastolic flow resulting in low RI. Arterialization of venous waveform.	• Confluent dysplastic blood vessels without associated soft tissue mass.	• Confluent dysplastic blood vessels without associated soft tissue mass. • Flow voids on spin echo sequences.	• Rapidly enhancing dysplastic blood vessels. • Early filling of hepatic and portal veins. • Transient hepatic attenuation/intensity differences (THAD/THID) secondary to arteriovenous shunting.

Table 2: Differential diagnosis table for liver mass in a newborn

ABBREVIATIONS

AFP: Alpha-fetoprotein
AV: Arteriovenous
AVM: Arteriovenous malformation
CT: Computed tomography
DMSO: Dimethyl sulfoxide
EVOH: Ethylene-vinyl alcohol copolymer
HB: Hepatoblastoma
HHT: Hereditary hemorrhagic telangiectasia
IHH: Infantile hepatic hemangioma
IVC: Inferior vena cava
MRI: Magnetic resonance imaging
NB: Neuroblastoma
RI: Resistive index
US: Ultrasound

KEYWORDS

Arteriovenous malformation; vascular malformation; liver; embolization; Onyx; interventional radiology