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VASCULAR MEDICINE

CASE REPORT: CLINICAL CASE

Complex Arterio-Portal-Venous Malformation of the Liver

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ABSTRACT

A neonatal female patient exhibited a congenital intricate vascular malformation affecting the liver, encompassing anomalies in the arterial, venous, and portal venous systems and notably including an aneurysm within the portal vein. The management strategy involved a staged endovascular approach, initially using retrograde embolization via the venous outflow tract. Subsequently, transarterial embolization was performed to address complications associated with pulmonary and portal hypertension. (J Am Coll Cardiol Case Rep 2024;29:102337) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

In the third trimester, a complex hepatic vascular malformation was diagnosed in utero of a female with known free trisomy 21 (47, XX, +21). Prenatal ultrasound revealed diffuse vascular infiltration of the entire liver with mixed flow, including arterioportal shunting and aneurysmal dilation of a large venous structure (**Figure 1A**). The child was delivered via cesarean section at 37+5 weeks with a birth weight of 3,020 g. Postnatally, the child experienced dyspnea, necessitating continuous positive airway pressure support in the pediatric intensive care unit.

LEARNING OBJECTIVES

- To understand the role of hepatic vascular malformations in Down syndrome.
- To dissect the different vascular territories involved in complex vascular malformations of the liver.
- To be able to tailor individualized endovascular treatment approaches to complex vascular malformations.

INVESTIGATIONS

Transthoracic echocardiography identified tricuspid insufficiency grade 2 with a gradient >60 mm Hg, corresponding to systemic pressure. Additional findings included patent ductus arteriosus, an atrial septal defect (ASD, type 2) with biphasic shunting, and a subaortic muscular ventricular septal defect.

Computed tomography angiography on day 1 confirmed a complex vascular malformation of the liver with multiple hypertrophic arterial feeders arising from the hepatic artery, phrenic artery, and an atypical internal thoracic artery, leading to massive arterioportal shunting. A large aneurysm of the portal vein with arterialized flow and patent ductus venosus was observed, covered only by the liver capsule and draining into the enlarged right hepatic vein

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ABBREVIATIONS AND ACRONYMS

ASD = atrial septal defect

EVOH = ethylene-vinyl alcohol copolymer

(Figures 1B to 1D). Notably, an extrahepatic portal vein could not be identified.

MANAGEMENT

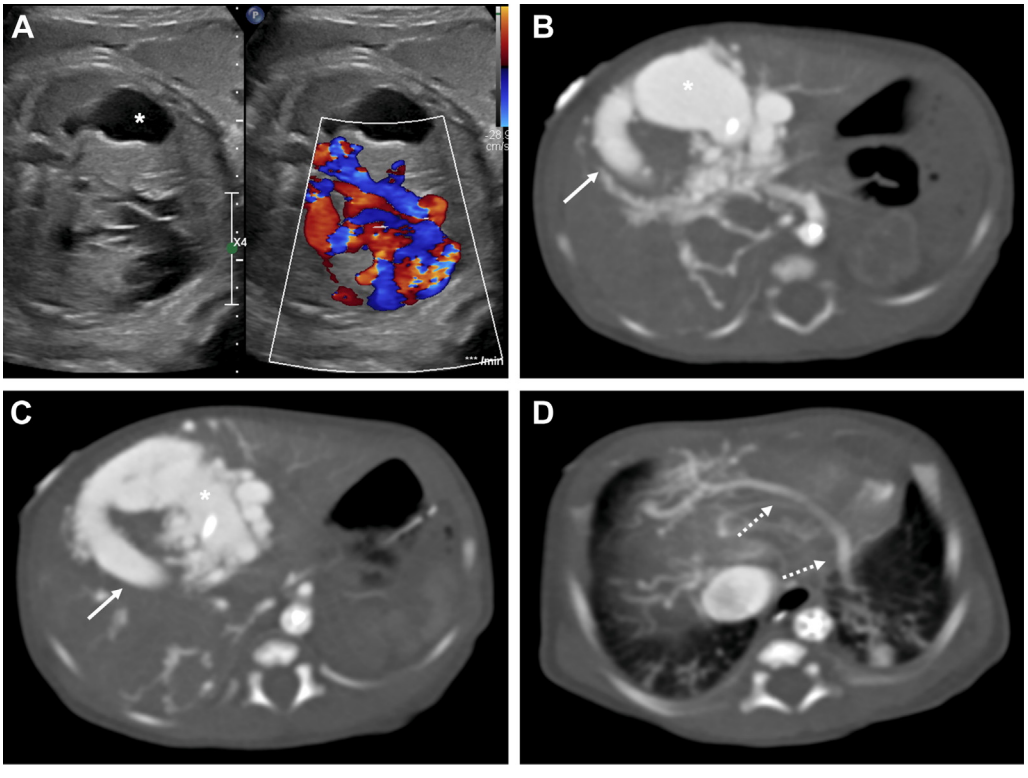
On day 2, cardiac catheterization confirmed systemic pressure in the right ventricle of 65 mm Hg and severe pulmonary hypertension. Subsequent access to the hypoplastic aorta was achieved via the patent ductus arteriosus followed by superselective angiography of the hepatic artery, confirming diffuse arterioportal shunting. Detachable microcoils were used to close various shunts (Figure 2), reducing the right ventricle pressure to 53 mm Hg.

During the subsequent course, oxygen saturation ranged from 75% to 80% when breathing room air and 90% when continuous positive airway pressure was supported by oxygen. Because of persistent massive arterioportal shunting, high-output cardiac failure, and massive pulmonary hypertension, cardiothoracic repair was deemed infeasible. The child was transferred to another hospital for subsequent treatment of the hepatic vascular malformation at 2 months of age.

Repeated computed tomography angiography revealed persistent arterialized perfusion of the intrahepatic aneurysm of the portal vein. The peripheral arterial vasculature for potential transarterial access presented substantially hypoplastic (Figure 3). Additionally, progressive hypoplasia of the abdominal aorta below the celiac trunk was observed (Figure 4). Yet, an atypical hypertrophic internal thoracic artery was present, leaving the retrosternal area at the level of the diaphragm, perforating the liver capsule, and feeding the intrahepatic vascular malformation.

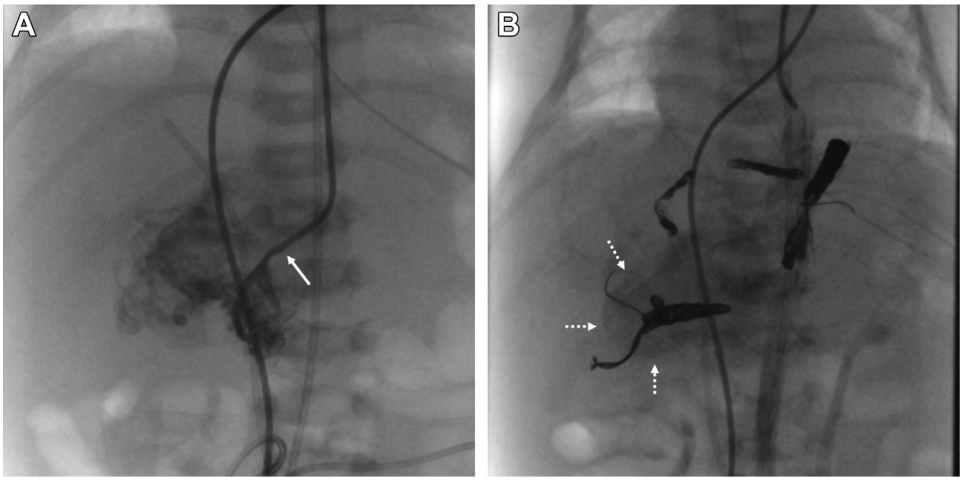
To reduce the left ventricle preload and prevent rupture of the subcapsular intrahepatic aneurysm, a

FIGURE 1 Initial Presentation of a Complex Hepatic Vascular Malformation



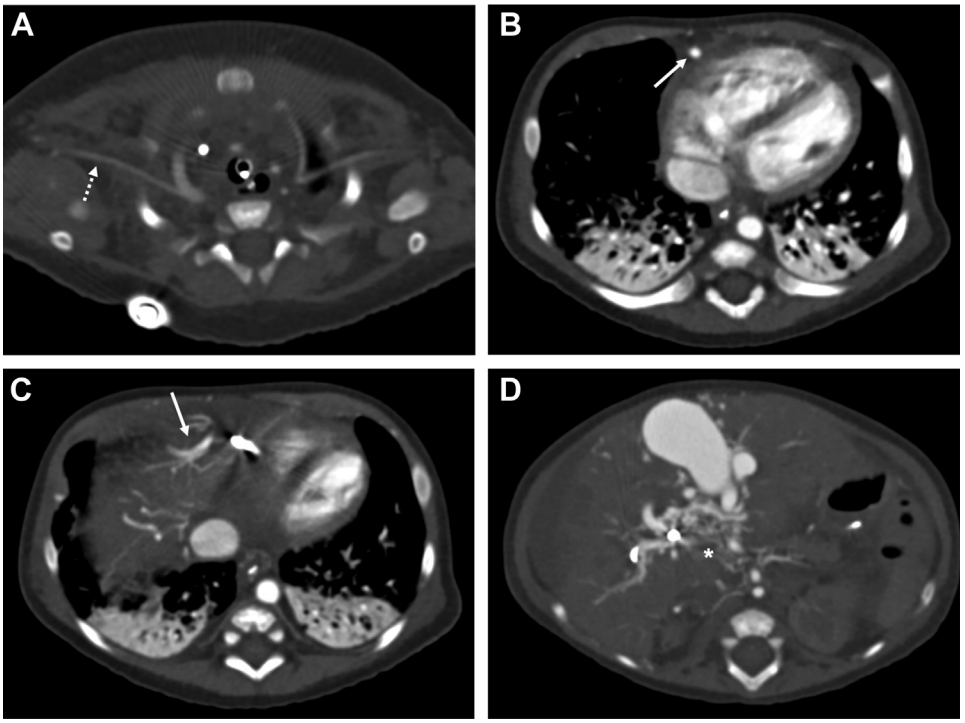
(A) Prenatal ultrasound in the third trimester revealed diffuse vascular infiltration of the liver parenchyma with mixed flow characteristics including an arterial component as well as aneurysmatic deformation of a major vessel (asterisk). (B and C) Computed tomography angiography obtained on day 1 after delivery revealed a large aneurysm of the portal vein (asterisk) with arterial contrast enhancement and a patent ductus venosus (arrow). (D) Multiple arterial feeders were identified including a major feeder originating from the phrenic artery (dotted arrow).

FIGURE 2 **Transarterial Angiography**



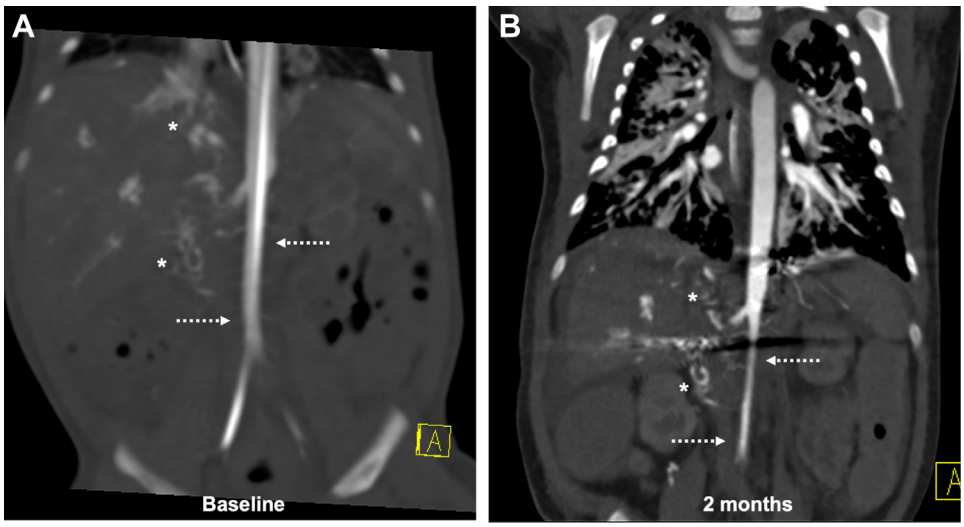
(A) Initial angiography obtained via a patent ductus arteriosus shows diffuse arterial feeders originating from the hepatic artery (arrow: catheter in the celiac trunk). (B) Major arterial feeders to the hepatic malformation from the hepatic artery were embolized using detachable microcoils, with the aneurysm of the portal vein still being perfused after embolization (dotted arrows).

FIGURE 3 **Computed Tomography Angiography for Planning of Subsequent Embolization**



(A) Computed tomography angiography revealed hypotrophy of the peripheral arterial tree with a diameter of the axillary artery of <0.7 mm. (B) A hypertrophic abnormal internal thoracic artery was present (2-mm diameter) perforating the liver capsule and (C) feeding the intrahepatic vascular malformation. (D) Persistent arterial perfusion of the portal venous aneurysm was noted with diffuse arterial feeders intrahepatic even after initial coil embolization.

FIGURE 4 Hypotrophy of the Abdominal Aorta

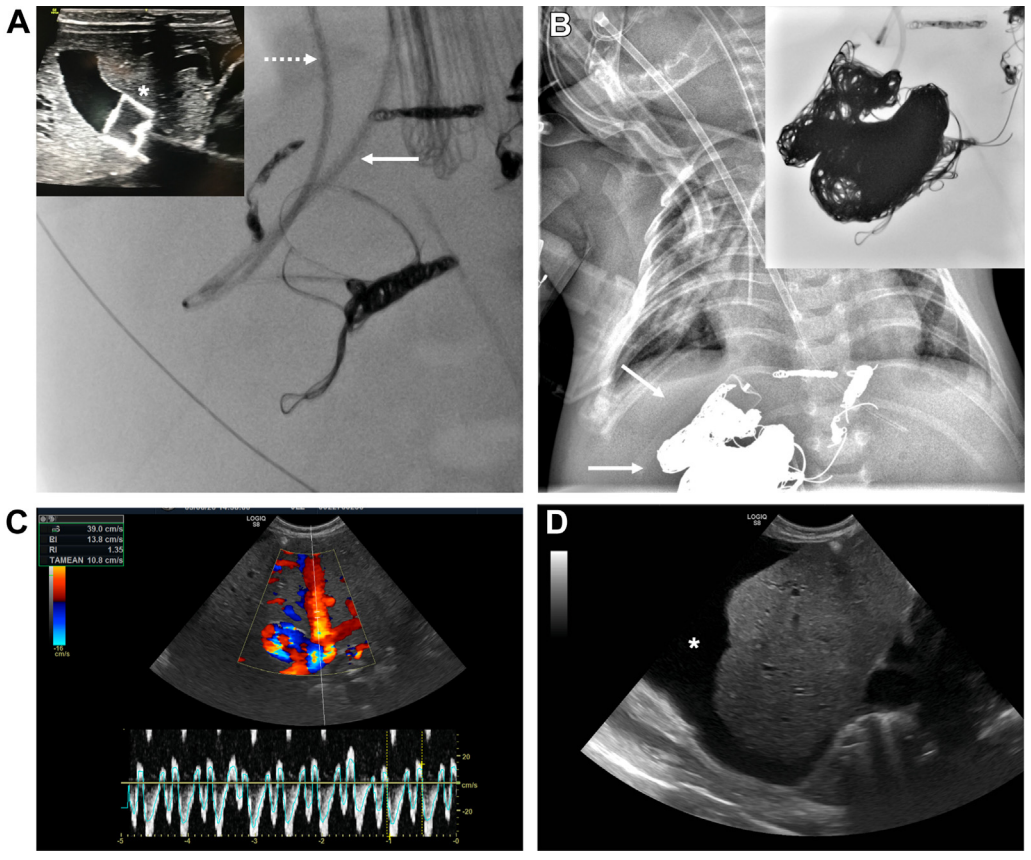


Comparison of the diameter of the abdominal aorta (dotted arrows) at (A) day 1 postdelivery compared to the (B) age of 2 months revealed substantial hypotrophy of the aorta below the origin of the celiac trunk attributed to a hepatic steal caused by massive intrahepatic shunting (asterisks).

transvenous retrograde embolization of portal vein aneurysm was performed next. A 4-F access was obtained on the left and a 6-F access via the right jugular vein. Via a 4-F catheter placed within the hepatic artery, a subsequent 2-F microcatheter was placed within the aneurysm. In order to prevent a coil dislocation from the liver, a type 2 Amplatzer Plug (Abbott Vascular) was unfolded temporarily (not detached) in the draining right hepatic vein in parallel to the 4-F catheter, which was removed after successful embolization with detachable microcoils (**Figures 5A and 5B**). After successful embolization of the intrahepatic aneurysm including the venous outflow of the vascular malformation, the pulmonary hypertension improved significantly with biphasic shunting via the ASD now being absent. However, obstruction of the venous outflow from the malformation with persisting arterial inflow (**Figure 5C**) resulted in conversion of the pulmonary hypertension to severe portal hypertension with ascites >450 mL/d (at a current weight of 3.6 kg) (**Figure 5D**). Arterialized blow flow, and more importantly arterialized pressure within the intrahepatic portal venous system, required additional embolization of the arterial inflow to the hepatic malformation.

As peripheral arteries were not amenable for transarterial access, a surgical cut-down was performed and the large internal thoracic artery was canulated surgically in the operating room. A 2.7F access was placed antegrade, pointing toward the hepatic malformation, and a 4F sheath was placed retrograde within the hypertrophic internal thoracic artery to access the aorta (**Figure 6A**). First, liquid embolization was performed using ethylene-vinyl alcohol copolymer (EVOH) via a dedicated microcatheter placed within the arterial feeders (**Figure 6B**). After successful closure, the abdominal aorta was catheterized via a 4-F catheter and subsequent superselective placement of another microcatheter in the hypertrophic phrenic feeders of the malformation, similarly being embolized with EVOH (**Figure 6C**), resulting in substantial devascularization of the arterial feeders (**Figure 6D**). Surgical removal of the sheaths and access closure was finally performed. Ascites production decreased significantly within the following days, allowing for removal of the ascites drain 14 days later, which was interpreted as a sign for decreased portal hypertension. The high cardiac output failure resolved and allowed for subsequent extubation, with the child requiring only 25% inspiratory O_2. Biphasic shunting remained absent, and the pulmonary hypertension was persistently relieved. Magnetic resonance imaging performed 14 days after embolization revealed a present

FIGURE 5 Retrograde Access for Embolization of the Portal Venous Aneurysm



(A) Bilateral transjugular access allowed for cannulation of the portal venous aneurysm with 1 catheter carrying the microcatheter for subsequent coil embolization (dotted arrow) and the second catheter (arrow) allowing for placement of a type 2 Amplatzer Vascular Plug to prevent coil dislocation to the systemic circulation (upper left image, asterisk). (B) Coil embolization led to complete occlusion of the portal venous aneurysm (arrows, magnification upper right panel). (C) Despite successful occlusion of the aneurysm, persistent arterial inflow into the hepatic malformation was observed afterward with the initial pulmonary hypertension being converted into (D) portal hypertension with large amounts of ascites (asterisk).

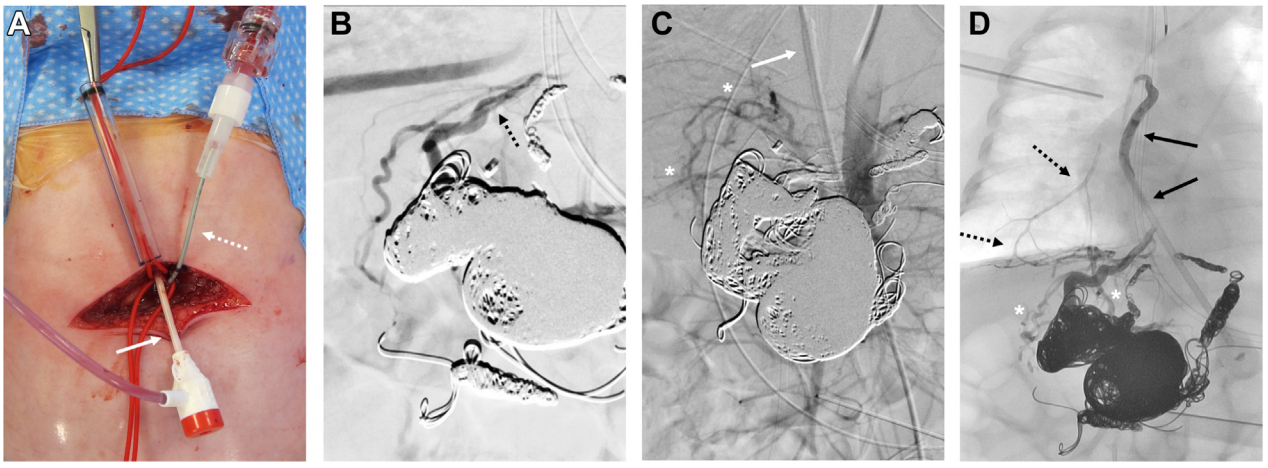
extrahepatic portal venous system and the aneurysm being completely occluded, with the reimaging malformation being subtotally devascularized (**Figure 7**).

DISCUSSION

Besides well-known cardiac anomalies in the setting of trisomy 21 (Down syndrome), rare congenital vascular malformations of the liver have been described. Especially when increased shunting across the liver is present, those malformations have been associated with cardiorespiratory failure.[1] Especially in trisomy 21, a large spectrum of clinical and anatomical features of portal vascular shunts has been reported but is always limited to series of a few cases only.[2] Because the clinical presentation and anatomy of the malformations is very heterogeneous, the therapeutic approach has to be tailored to the severity of the symptoms.

For proper management, a systematic assessment of complex vascular malformations is required. First, it has to be clarified which vascular compartments are involved, which in case of hepatic malformations can be the arterial system, the portal venous system including the ductus venosus (patent or occluded), and the hepatic venous system. For complex malformations, cross-sectional imaging may be required to dissect the components of the hepatic vasculature. The type and degree of shunting can be estimated by ultrasound. Although in most cases the liver parenchyma appears normal and liver function is preserved, both shunting as well as closure of shunts can

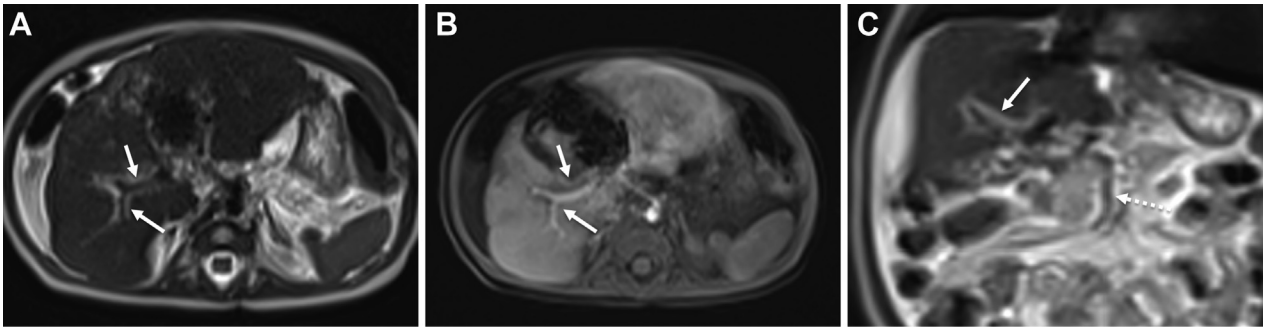
FIGURE 6 Transarterial Access and Embolization of the Arterial Component



(A) Surgical cut down to the abnormal internal thoracic artery was performed with open cannulation of the artery antegrade (2.7-F, dotted arrow) and retrograde (4-F, arrow). (B) First antegrade embolization of the arterial feeder was performed using ethylene-vinyl alcohol copolymer (dotted arrow). (C) Secondly, the aorta was catheterized in a retrograde manner (arrow) with subsequent superselective embolization of arterial feeders via the hypertrophic phrenic artery (asterisk), resulting in (D) subtotal devascularization of the arterial inflow to the hepatic vascular malformation with feeders from the abnormal internal thoracic artery being occluded (dotted arrows) and the artery itself (arrows) allowing for safe removal of the sheaths as well as the feeders from the phrenic artery (asterisks).

result in a deterioration of liver function because of altered perfusion characteristics. Endovascular treatment has emerged as first-line treatment, especially in critically ill patients in which surgery is associated with increased risk. In our case, the arterialized blood flow within the large aneurysm of the portal vein with minimal tissue coverage together with the impaired cardiopulmonary function made a surgical approach such as hemihepatectomy or liver transplantation impossible. Embolization of hepatic vascular fistulas and malformations is well established and can be performed using both coils and liquid embolics.[2,3] Especially in cases with shunting from the portal vein, the retrograde approach has been described as safe and effective.[4] However, in our case, the malformation was not limited to a portosystemic shunt only but included complex arterial inflow feeding an aneurysm of the portal vein, requiring access via different vascular territories in a staged approach. Being able to reduce the pulmonary hypertension by limiting the hepatic outflow from the large arterialized aneurysm, the

FIGURE 7 MRI Showing the Presence of an Intrahepatic and Extrahepatic Portal Venous System



(A) T2-weighted magnetic resonance imaging (MRI) showing a present intrahepatic portal vein stem on the right with (B) perfusion after contrast administration. (C) Regular draining of the superior mesenteric vein (dotted arrow) toward the portal venous system (arrow) on T2-weighted MRI.

embolization resulted in a conversion of pulmonary to portal hypertension. With liquid embolic agents being regarded as superior in the management of arteriovenous malformations,[5] we opted for embolization of the diffuse arterial feeding network using EVOH. The resulting decreased portal hypertension allowed for subsequent cardiothoracic surgery and repair of the large ASD. After successful endovascular treatment of the complex hepatic vascular malformation, an intact extrahepatic portal venous system was present. Therefore, liver transplantation as a potential cure of the hepatic disease manifestation was discussed. Although liver transplantation is routinely feasible in newborns, reports in the setting of Down syndrome are limited to a few cases described worldwide.[6] Because of cardiopulmonary comorbidities and impaired immune status, liver transplantation was not considered reasonable in this case. In due course, the child suffered from progressive intestinal malabsorption, frequently being present in Down syndrome,[7] but potentially being aggravated by increasing portal hypertension caused by the recurrence of arterial inflow to the hepatic malformation.

FOLLOW-UP

Nutrition of the child was subsequently completely oral, and the child started to gain weight. After another 2 months, open cardiothoracic repair of the ASD was performed, with the child recovering well and allowing for discharge at the age of 6 months.

CONCLUSIONS

The incidence of hepatic vascular malformations is notably higher in the pediatric population diagnosed with Down syndrome. These malformations have the potential to affect all vascular compartments within the liver. The goal of endovascular treatment is to reduce shunt flow and occlude associated aneurysms. In cases involving complex vascular anomalies, staged treatments may be necessary and can lead to improved cardiopulmonary function. Continuous assessment of the comorbidities associated with this complex syndrome is crucial because these factors can significantly impact the overall outcome in affected pediatric patients.

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