Successful embolization of a congenital intra-hepatic arterioporal fistula in a neonate with the MVP Microvascular Plug system (MVP-3Q)

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**Abstract**

This case report describes a neonate with an antenatally diagnosed vascular anomaly of the liver. Ultrasound at birth confirmed an arterioporal fistula communicating the left hepatic artery and an anterior branch of the right portal vein. Computer tomography angiography on day 7 of life redemonstrated the arterioporal fistula and defined the vascular anatomy for potential treatment. Transarterial embolization of the arterioporal fistula was performed at 3 weeks of life using an MVP Microvascular Plug System 3Q (Reverse Medical Corp, Irvine, CA, USA). Intra-procedural angiography showed successful occlusion of the fistula, patency of the portal vein with hepatopetal flow, and patency of the hepatic artery with no signs of arterial or venous thrombosis. There were no intra- or post-procedure complications. Multiple follow-up ultrasounds at 1-13 months showed stable occlusion of the embolized fistula with no evidence of recanalization, with the patient having a normal life and no sequelae. This case illustrates a successful novel approach to manage the rare condition of a solitary hepatic arterioporal fistula in a neonate using the MVP system. Current literature on congenital arterioporal fistulas and the MVP system is reviewed.

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**Keywords:**
Arterioporal fistula
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**Abbreviations:** IAPF, intrahepatic arterioporal fistula; US, ultrasound; MVP, Micro Vascular Plug; CT, computed tomography; HA, hepatic artery; TAE, transarterial embolization; PTFE, polytetrafluoroethylene; CHM, congenital heart malformation.

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Introduction

Congenital intrahepatic arteriportal fistula (IAPF) refers to a rare intrahepatic vascular anomaly connecting the hepatic artery to the portal vein [1]. IAPFs can be asymptomatic or present in infancy with symptoms of portal hypertension and rarely congestive heart failure, warranting prompt recognition and treatment [2]. Doppler ultrasound (US) can detect IAPFs in neonates and infants [2]. Currently, transarterial embolization is the first-line treatment for IAPFs as it is minimally invasive and shows favourable outcomes for fistula obliteration [3]. This report illustrates the case of a neonate with an antenatally diagnosed IAPF confirmed by US at birth and treated with transarterial embolization at 3 weeks of life using the MVP Microvascular Plug system (Reverse Medical Corp, Irvine, CA, USA). Although other reports have described interventional treatment of IAPFs, to the best of our knowledge, this is the first case that uses the MVP system for embolization of an IAPF in a neonate.

Case report

Informed consent was obtained from the patient’s parents for this publication. This report describes a neonate who was antenatally diagnosed with an arteriovenous anomaly of the liver at 20 weeks gestational age via obstetric US. Transabdominal US on day 1 of life confirmed a lesion located in the upper central portion of the liver, characterized by a vascular saccular structure with an arterial feeder arising from an enlarged left hepatic artery and a draining venous channel connecting with the right portal vein, consistent with an arteriportal fistula (Fig. 1). The right hepatic artery was not visualized on US. The portal vein showed hepatopetal flow. The neonate had no symptoms concerning for high output cardiac failure secondary to the vascular anomaly, with an echocardiogram demonstrating normal biventricular systolic function, a patent foramen ovale shunting left to right, and a small patent ductus arteriosus shunting bidirectionally that later closed. There were no clinical or imaging signs of portal hypertension. Computed tomography (CT) in arterial and portal venous phase (with 3D reformats) of the abdomen at 7 days of life re-demonstrated the large intrahepatic solitary fistula between the enlarged left hepatic artery and an anterior branch of the right portal vein with an associated saccular structure extending anteriorly to segments IVA and VIII (Fig. 2). A small patent right hepatic artery was seen. There was no connection of the fistula with the hepatic venous system. The aorta was noted to have a decreased caliber below the level of the fistula, measuring 5.7 mm superior to the celiac axis origin and 3.1 mm just inferior to the origin of the renal arteries (Fig. 2). Liver enzymes and tests of synthetic function were normal, with the exception of a mildly elevated aspartate aminotransferase. A multidisciplinary decision between treating teams was made to perform embolization of the IAPF to prevent growth of the lesion, which could potentially compromise the clinical condition of the patient. It was also agreed to wait until week 3 of life to make the procedure safer in terms of vessel caliber. While waiting for the procedure, the patient developed urosepsis on day 11 of life and was treated with a 2-week course of antibiotics.

At 3 weeks of life, weighing 4.3 kg, the patient presented for image-guided embolization of the IAPF. The pre-procedure US showed a mildly decreased caliber of the saccular structure compared to previous imaging and no free fluid (Fig. 3). The procedure was performed under general anesthesia and sterile conditions. The right brachial artery was used for access as it allowed for easier entry into the celiac trunk/hepatic artery and prevented instrumentation of the narrowed lower abdominal aorta. Access was obtained into the proximal brachial artery using US guidance and a 4 French vascular sheath was placed. A total of 100 U/kg (430 units) of heparin were given by the anesthesiologist as per institutional protocol. Forty-five micrograms of nitroglycerin divided into 3 injections were delivered via the vascular sheath during the procedure. The common hepatic artery was then catheterized with a 4 French Berenstein catheter and a 0.035” guidewire. A Renegade microcatheter was used for super-selective catheterization of the left hepatic artery. Intra-procedure angiograms re-demonstrated the known vascular anomaly and the presence of a single arterial feeding channel with a relatively straight course, (measuring 12 mm in length and 2 mm in diameter) making

Fig. 1 – Gray-scale and color-Doppler US on day 1 of life showing: (A) Prominent celiac trunk (arrow) and an enlarged common hepatic artery (arrowhead). (B) Saccular structure with smooth margins (arrows) located in the central superior portion of the liver and in contact with the anterior liver capsule. (C) Prominent vascular flow within the saccular structure was observed, with aliasing artifact reflecting turbulent flow. (D) Very tortuous and prominent left hepatic artery (arrow) which originated the arterial feeder of the vascular anomaly. (E) Vascular channel (white arrow) communicating with the right anterior branch of the right portal vein (black arrows). This constellation of findings was compatible with a congenital intrahepatic arteriportal fistula.
Fig. 2 – An enhanced CT of the abdomen was performed to better characterized the lesion. (A) Sagittal MIP in arterial phase showing a prominent celiac trunk (black arrow) and enlarged common hepatic artery (black arrowhead). The abdominal aorta below this area showed decreased caliber (white arrows). (B) Axial MIP reformat demonstrating the arterioportal fistula involving segments IVA and VIII. It comprises the arterial feeder (arrowhead), the saccular structure (black arrows), and the portal communicating venous channel (asterisk). The right liver parenchyma showed an earlier opacification with contrast, most likely secondary to the fistula. (C) Axial MIP reformating the origin of the arterial feeder from the left hepatic artery (arrowheads) and the junction of the portal communicating venous channel with a branch of the right portal vein (black arrows). (D) Sagittal MIP reformating the straight portion of the arterial feeder (arrows) and its junction with the saccular structure (arrowhead). The straight trajectory of this portion allowed the deployment of the MVP. (E, F) 3D reconstructions showing the large saccular structure (S), the left hepatic artery (HA) giving rise to the arterial feeder (white arrows) and the draining venous channel to the portal vein (P).

Fig. 3 – Intra-procedural findings. (A) Gray-scale US pre-embolization showing a mildly decreased caliber of the saccular structure (S) and the straight portion of its arterial feeder (arrows). (B, C, D) Intra-procedure super selective arteriograms in different projections, with the microcatheter positioned at the arterial feeder, showing anatomical details of the arterioportal fistula characterized by the straight portion of the arterial feeder (white arrow), the saccular structure (S) and the draining venous channel (black arrow, P). Satisfactory peripheral portal venous flow was observed (black arrowheads). Short branches were observed from the saccular structure (white arrowhead). (E) Deployment of the MVP within the straight portion of the arterial feeder via the microcatheter. The distal (black arrow) and proximal (white arrow) radiopaque markers were clearly visualized, in an adequate position as determined by previous planning angiograms. The device was deployed with no incidents. (F) Control angiogram 10 minutes after deployment of the MVP (black arrow) demonstrated successful occlusion of the fistula. There was no opacification of the saccular structure or portal branches.

it technically feasible to deploy a plug (Fig. 3). Prophylactic antibiotic was given. A Micro Vascular Plug (MVP-3Q, 5.3 mm diameter - 12 mm length - vessel size 1.5-3.0 mm) was deployed under fluoroscopic guidance within the straight portion of the arterial feeder, proximal to the saccular structure and slightly distal to the main trunk of the left hepatic artery (Fig. 3). An angiogram 10 minutes post MVP deployment showed satisfactory fistula occlusion (Fig. 3) and adequate opacification of the common hepatic artery and its branches and abdominal aorta. US of the liver confirmed successful fistula occlusion and patency of the portal vein with hepatopetal flow. The brachial sheath was then removed and hemostasis was achieved by manual compression. The right arm showed a minimal decrease in temperature and a mild decrease in distal capillary refill after sheath removal. The thrombosis team advised that there was no indication for prophylactic anticoagulation as the blood flow in the right portal vein would be sufficient to prevent stasis.

Follow-up US at 1, 4, 8, and 13 months showed stable appearance of the liver at the site of the IAPF with no evidence of recanalization (Fig. 4). After a few months, the saccular structure was no longer recognizable (Fig. 4). There were no abnormalities or sequelae of the right arm. Genetic testing for hereditary hemorrhagic telangiectasia returned negative. The patient was discharged from care at 13 months with appropriate growth and development for the age.

Discussion

IAPF is an abnormal vascular connection between the hepatic artery and portal vein [1]. IAPF is a rare occurrence, with literature describing less than 50 cases to date [4]. IAPFs can be congenital (primary) or acquired (secondary) [2]. Acquired IAPFs are more common and arise following events such as
blunt or penetrating liver trauma, hepatic neoplasms, hemangiomas or hepatic artery aneurysms, fine needle liver biopsy, hepatectomy, and percutaneous transhepatic biliary drainage [2,5]. Congenital IAPFs involving the hepatic artery occur in less than 10% of cases [6]. A few classification systems of IAPFs have been reported, based on either location or vascular supply of the fistula [2,7]. Norton et al. proposed 3 subtypes of IAPFs: unilateral (type 1) IAPF supplied by only one of the right, left or main hepatic arteries (HA); bilateral (type 2) IAPF supplied by both HAs; and complex (type 3) IAPFs supplied by a vascular plexus with several feeding arteries [2]. About 75% of congenital IAPFs present by 2 years of age with clinical manifestations of portal hypertension, including abdominal distension, gastrointestinal bleeding, diarrhea, and failure to thrive [2,3,8]. In rare cases where the ductus arteriosus remains patent, high-output congestive heart failure can occur [2]. Congenital IAPFs are commonly an isolated finding, but can be associated with syndromes such as hereditary hemorrhagic telangiectasia, Ehlers-Danlos or Trisomy 21 [3,5]. Diagnosis of IAPF usually relies on color-Doppler US, which may show hepatic artery enlargement, pulsatile hepatofugal flow in the portal vein, and portal vein dilatation [3]. Contrast-enhanced CT and contrast-enhanced magnetic resonance imaging may be used to confirm the US findings [3]. Conventional angiography is the gold-standard imaging modality as it can precisely outline the vascular anatomy and also serve therapeutic purposes [9]. Management of IAPF focuses on occlusion of the fistula and recovery of normal intrahepatic hemodynamics [2]. To date, there are no published cases of spontaneous IAPF closure in children [9]. Therapeutic options include (1) transarterial embolization (TAE); (2) surgical approaches such as hepatic artery ligation or partial hepatectomy; or (3) combination of TAE and surgery [2]. The choice of treatment depends on the location and size of the fistula, as well as hospital resources and expertise [2]. TAE is the preferred therapeutic option as it decreases morbidity and post-procedural pain, minimizes hospital stay, and carries lower costs compared to surgery [2].

A variety of embolic agents are available for embolization such as coils, sclerosing agents, covered stents, and vascular plugs. The choice of agent depends on the location and size of the fistula and local expertise [4]. The MVP is a deployable embolic device composed of an expanding nitinol frame coated with impermeable polytetrafluoroethylene (PTFE), attached to a guidewire and delivered through a catheter or microcatheter (according to the MVP size) [10]. There are currently 4 models of MVPs that differ in their diameter (3, 5, 7 and 9 mm), each used for vessels of different caliber. MVP-3 and MVP-5 are deployed via a microcatheter while MVP-7 and MVP-9 employ a 4 French or 5 French diagnostic catheter [10]. In our case, the MVP-3Q was used. MVP offers several advantages over other embolic agents like coils. In particular, (1) compatibility with most common catheters and microcatheters; (2) navigability through tortuous vessel anatomy given the small size and flexible skeleton; (3) potential for easy repositioning without device or vessel disruption; (4) instant target vessel occlusion; and (5) stability of plug without migration [11]. Specifically regarding coils – which may require the use of multiple devices to occlude an artery – MVP differs by the fact that it can usually be used as a single device, as seen in this case [11].

MVP use has been described in the adult population for numerous applications, including hepatic artery embolization [10]. One of the earliest reports of MVP use describes embolization of the gastroduodenal artery in a 7-year-old male [12]. There are also scattered reports illustrating MVP use for vascular embolization in children with congenital heart malformations [CHM]. One recent review described successful embolization of abnormal veno-venous and aorto-pulmonary connections in ten patients (median age 3 years) with CHM; all cases were free of complications [13]. However, to the best of our knowledge, this is the first report of MVP use for IAPF embolization in a neonate. One case report described embolization of a large IAPF in a 13-year-old boy using an Amplatzer occlusion device, which uses a similar plug principle as the MVP [14]. However, it is larger, lacks the PTFE coating, requires a stiff delivery cable and uses a diagnostic instead of a microcatheter, which makes it challenging to use in small children [13–15]. Another case report discussed embolization of IAPF in an 11-day old male presenting with cardiac failure using micro Nester coils via a direct transhepatic approach [16].

The majority of published reports in children describe a transfemoral arterial or transhepatic portal venous access for embolization [16]. In the adult population, transradial arterial access is gaining momentum due to lower bleeding and vascular complications, quicker recovery times, and lower costs compared to transfemoral access; but the smaller size of the
radial artery in children may make it technically challenging in the pediatric population [17,18]. Transaxillary access is also an established procedure in adults and has been reported by Roebuck et al. to be successful for arteriography purposes in 19 children aged between 7 days and 15 years [18]. The current case was completed through a right transbrachial arterial access. However, the left side is typically preferred to the right due to easier entry into the descending aorta and as it involves intersecting the root of only 1 cerebral arterial [18]. The brachial artery is generally not recommended for use due to its proximity to the median nerve, risk of thrombosis and vasospasm with subsequent limb ischemia [19,20]. However, literature on brachial artery catheterization in neonates is scarce. Schindler et al. reported 112 cases of successful brachial artery catheterization for hemodynamic monitoring with few complications including temporary occlusion (n = 1), local infection (n = 1), and local hematoma (n = 5) [20]. US guidance is recommended for access as it can increase catheterization success rates in children [21]. Nitroglycerin – a smooth muscle relaxant that results in arterial and venous dilatation – can be used to mitigate the risk of vasospasm and limb ischemia [19]. Intravenous nitroglycerin was used in our case; however, topical nitroglycerin use has also been described [19]. The infant needs to be carefully monitored when using nitroglycerin due to its side effects of hypotension, tachycardia, and methemoglobinemia due to nitric oxide production [19].

In conclusion, the present case illustrates the successful use of the MVP system in treating a neonate with a congenital IAPF. The MVP can be an effective embolic agent that has the benefit of using a microcatheter and allows easier navigation through tortuous vessel anatomy. Larger studies or case series are needed to evaluate the safety and efficacy of this device in the pediatric population for treating gastrointestinal vascular malformations or in other applications.

Patient consent

Written informed consent was obtained from the patient’s parents (legal guardians) for publication of this case report and accompanying images. This consent was attached to the patient’s chart.

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