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Case Report

The Congenital Intrahepatic Arterioportal Fistula Syndrome: Elucidation and Proposed Classification

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ABSTRACT
Congenital intrahepatic arterioportal fistula is a rare but treatable cause of portal hypertension for which early recognition may lead to successful radiological management. We report an infant presenting with severe failure to thrive, melena and splenomegaly due to a congenital intrahepatic arterioportal fistula, successfully ablated after multiple trials of superselective transarterial embolization. Comprehensive review of congenital cases provides an understanding of the key clinical features defining this syndrome. A classification system is proposed, upon which treatment decisions may be based. ***JPGN 43:248–255, 2006.*** **Key Words:** Arterioportal fistula—Portal hypertension—Malabsorption—Gastrointestinal hemorrhage—Embolization—Pediatrics—Congenital
Abbreviations: AVM, arteriovenous malformation—CHF, congestive heart failure—GI, gastrointestinal—HA, hepatic artery—IAPF, intrahepatic arterioportal fistula(e)—PV, portal vein—TAE, transarterial embolization—US, ultrasound.

CASE PRESENTATION

A white boy was hospitalized at 2-years-old for evaluation of abdominal pain, melena and profound failure to thrive since 6 months. He was born at term weighing 2840 g after an unremarkable pregnancy. No umbilical catheterization had been performed, and there was no history of any abdominal trauma. There was no family history of note. He was breast-fed and thrived for his first 6 months. Between 6 months and 1 year of age, his weight fell from the 25th to below the 3rd percentile and his length fell below the 10th percentile. He developed abdominal distension, loose stools, muscle wasting, pallor and fatigue. His hemoglobin was 54 g/L (normal, 107–131 g/L) and ferritin 14 μg/L (normal, 6–30 μg/L). Antiendomysial and antitissue transglutaminase antibodies were negative, and sweat chloride testing was normal. His stools were intermittently positive for occult blood. Small bowel biopsies demonstrated subtotal villous atrophy with a mild lymphoplasmacytic infiltrate but no other features of celiac disease. Results of 72-hour fecal fat collection, serum immunoglobulin E by radioallergosorbent testing to cow's milk and soy protein, small bowel barium series and Meckel scan were normal.

Between 13 months and 2 years of age, his head circumference percentile declined from the 25th to below the 3rd percentile. He developed macrocytic anemia with hemoglobin of 80 g/L and mean corpuscular volume of 94.9 (normal, 70–86 fL). Red blood cell folate and serum vitamin B_{12} were normal. At 24 months, he presented with melena, intermittent abdominal pain with distension and nonbilious, nonbloody emesis. Physical examination revealed a pale thin child with distended abdomen, splenomegaly, ascites and hydroceles. Prominent distended superficial veins were noted in the anterior chest and abdominal wall; their filling below the umbilicus was cephalad. No hepatic bruit or venous hum was detected. Total protein and albumin were reduced at 47 and 25 g/L, respectively, whilst white blood cell and platelet counts, clotting profile, fibrinogen and liver biochemistry were

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normal. Upper endoscopy revealed esophageal varices without signs of recent bleeding and prominent submucosal vessels in the gastric corpus, antrum and proximal duodenum. No fresh or altered blood was seen in the proximal gastrointestinal (GI) tract. Histopathology demonstrated numerous capillary-sized ectatic vascular channels in the gastric lamina propria extending to nearly surface epithelium.

At 26 months, Doppler abdominal ultrasound (US) suggested a fistula between the hepatic artery (HA) and the portal vein (PV) in the left lobe of the liver, with reversal of portal flow, arterialized portal waveform, splenomegaly and ascites. Intravenous contrast-enhanced computed tomography (CT) revealed a prominent celiac axis, splenomegaly, ascites and small bowel wall thickening. Three-dimensional CT scan reconstruction delineated the vascular anatomy (Fig. 1). Angiography confirmed an intrahepatic arterioportal fistula (IAPF) with diffuse connections between intrahepatic branches of hepatic arteries and portal veins (Fig. 2), best defined in the left lobe. Arterial embolization was carried out via left-sided connections using 3 coils, ablating the fistula. Left portal venous pressure was no longer pulsatile and portal venous flow became hepatopetal. Post-procedure US showed normal antegrade flow in the main and left portal vein. The child was discharged completely well 5 days post procedure.

Intermittent melena recurred several months later. Doppler US showed appropriate flow through the portal venous system. At 30 months, endoscopy with biopsies was unchanged and no site of bleeding was identified. At 34 months, he was admitted pale, lethargic and wasted. He required multiple blood transfusions after octreotide injections were unsuccessful. US demonstrated pulsatile flow through right and left portal veins, enlarged hepatic arteries suggestive of arterioportal shunting and splenomegaly. Angiography demonstrated enlargement of the common, right and left hepatic arteries and widespread peripheral fistulae between HA and PV branches throughout both liver lobes. After

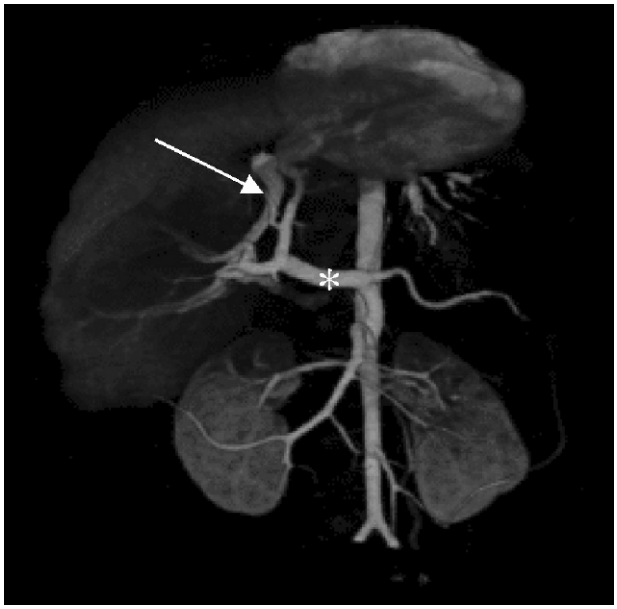

FIG. 1. CT angiogram with 3-dimensional reconstruction of child with IAPF at 2 years of age. Markedly enlarged common hepatic artery (asterisk) and left hepatic artery are observed. Early abnormal filling of the left branch of the portal vein is observed (arrow).

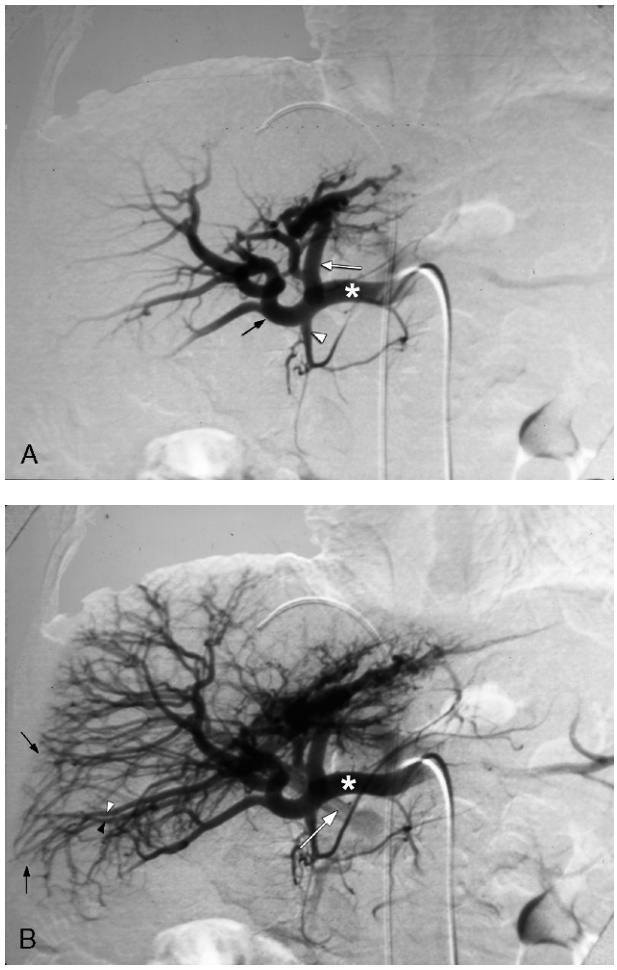

FIG. 2. Pretreatment hepatic arteriogram of child with IAPF at 2 years of age. A, Early phase of preprocedural hepatic arteriogram with selective catheter injection of an enlarged common HA (asterisk) shows early origin of left hepatic artery (white arrow) and disorganized vasculature in the left lobe of the liver. Lack of arborisation in this lobe is evident, suggestive of vascular "steal" from peripheral parenchymal arterial supply. The right hepatic artery (black arrow) and gastroduodenal artery (white arrowhead) are also indicated. Vascular pattern in the right lobe of the liver is normal. B, Hepatic arteriogram (late phase, image from same study as in A with 2-second delay) with catheter in common hepatic artery (white asterisk) demonstrates numerous widespread peripheral arterioportal fistulae (large black arrows) between hepatic artery branches (white arrowhead) and portal vein branches (black arrowhead) in the right and left liver. Portal vein branches parallel hepatic artery branches. There is rapid filling through the fistulae into the opacified main portal vein (large white arrow). Arterial morphology in the left liver is disorganized.

selective right HA catheterization, multiple microcoil embolizations were undertaken with satisfactory occlusion, with the exception of a residual fistula between a right HA branch and the left PV. GI bleeding resolved. At discharge, his hemoglobin was stable at 91 g/L. Abdominal US however showed persistent communication between a branch of the right HA and the left PV. Symptoms improved and his weight increased (from below the 3rd to the 3rd percentile).

At 38 months, melena recurred, accompanied by emesis, watery diarrhea, abdominal distension, increased splenomegaly and anemia. White blood cell and platelet counts were reduced, suggesting that hypersplenism had developed. Angiography demonstrated diffuse arterioportal shunting. Three anterior-posterior divisions of the right HA and one branch of the left HA were embolized with *N*-butyl 2-cyanoacrylate glue (Fig. 3), leading to significant reduction in shunting. However, slight residual shunting in the left lobe was evident from an accessory left HA and left gastric artery.

At 5 years of age, 3 years after initial embolization, the child was well, demonstrating good catch-up growth with resolution of symptoms, distension and ascites. No evidence of bile duct injury was seen. Abdominal US demonstrated normal portal flow with no evidence of portal venous hypertension or thrombosis.

DISCUSSION

Although some previous reviews (1,2) have included pediatric cases of intrahepatic arterioportal fistula, none are devoted to the congenital form of IAPF based on a comprehensive set of published cases. Moreover, none have fully characterized or introduced a classification system for this congenital syndrome.

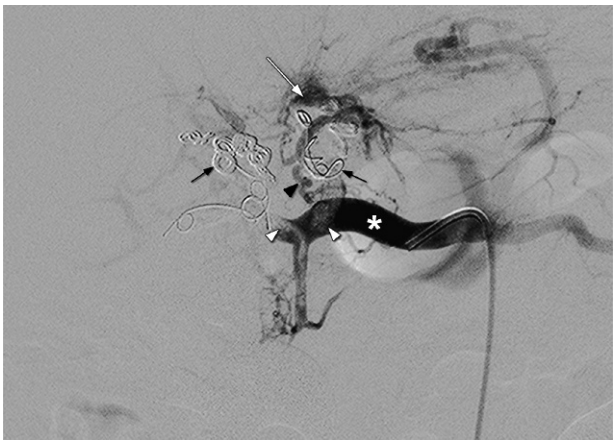

FIG. 3. Hepatic arteriogram of child with IAPF at 3 years of age after last treatment. After final glue and coil embolizations, hepatic arteriogram with catheter in enlarged common hepatic artery (white asterisk) shows embolization coils in right and left hepatic arterial branches from 2 previous catheter coil embolization procedures (black arrows). Residual glue is indicated in common hepatic artery and right hepatic artery (white arrowheads). Reduction in arterial flow throughout the liver, including the area of previously disorganized vasculature (white arrow), is observed after glue embolization. A small hepatic arterial branch in the left liver remains patent (black arrowhead).

Congenital IAPF, or hepatoportal fistula, is a rare cause of portal hypertension (PH) and gastrointestinal hemorrhage in infancy and early childhood. We propose to define congenital IAPF as an intrahepatic communication between the systemic, usually hepatic, arterial system and the portal venous system, without any communication with the systemic venous circulation, secondary cause or primary hepatic or biliary disease, presenting before 18 years of age. Increased blood flow in the portal system leads to presinusoidal PH (3).

Diffuse, or multiple, IAPF are virtually always congenital in origin (4,5), whereas a solitary fistula is typically acquired (6). Less than 10% of all arterioportal fistulae that involve the hepatic artery are congenital (1). Secondary causes of IAPF are more common and well described, including major blunt (7,8) or penetrating (9–11) abdominal trauma, surgical procedures such as needle liver biopsy (12–14), Kasai portoenterostomy (15–17) and segmental liver transplantation (18), hepatic artery aneurysm (14,19), cirrhosis (20–22), hepatocellular carcinoma (23–25), biliary atresia (26) and hereditary hemorrhagic telangiectasia (27–30).

Fistulae identified in adulthood are difficult to establish as congenital (4). The present case was considered to be congenital because of its symptomatic presentation in infancy, multiple fistulae, lack of any secondary factors and consistency with relevant reported cases.

Proposed Classification

An angiographic classification of congenital IAPF is proposed in Table 1. The classification system was derived from detailed analysis of original case reports of congenital IAPF published in the international literature. Case reports were obtained using a systematic search strategy. Our classification system is based primarily on the observations that therapeutic occlusion is normally directed at the arterial side and that outcome after various treatments is heavily influenced by the arterial vascular anatomy. We have classified IAPF according to the afferent vessels supplying them—unilateral, bilateral or complex. A unilateral IAPF (type 1) is supplied by only one of the right, left or main hepatic artery. Bilateral lesions (type 2) include supply from both of the parent hepatic arteries or their branches. Complex lesions (type 3), as in the present case, consist typically of a plexiform vascular nidus with multiple feeding arteries, *including* supply from arteries *other* than the hepatic arteries (eg, gastric artery).

As we encountered, new fistulae may become recognizable after initial embolization of larger fistulae, or existing fistulae may not be visualized, so the very

TABLE 1. *Proposed "Norton-Jacobson" Classification of Congenital IAPF*

Type	Afferent Supply	Description Of Afferent Vessel(s)	Initial Treatment
1	Unilateral (U)	RHA or LHA or main HA only	TAE
2	Bilateral (B)	Both RHA and LHA* or equivalent supply from main HA†	TAE Ligation if TAE fails
3	Complex (C)	RHA and/or LHA *and* nonhepatic arterial supply‡	Ligation/surgery§

*Both parent hepatic arteries and/or branches from both parent arteries.

†Equivalent supply from the main hepatic artery may take the place of one of the parent arteries.

‡Examples of other arteries include celiac, superior mesenteric, gastric, gastroduodenal, inferior phrenic, pancreaticoduodenal, jejunal, cystic, adrenal and splenic arteries.

§The current case was the first case of congenital IAPF treated exclusively by TAE; thus, data are insufficient to recommend TAE as initial therapy at this time.

IAPF, intrahepatic arterioportal fistula; RHA, right hepatic artery; LHA, left hepatic artery; HA, hepatic artery; TAE, transarterial embolization(s).

first angiographic appearance will not always properly classify the fistula type. The classification system therefore carries this limitation. However, the situation of one type "evolving" into another has arisen in less than one fifth of reported cases, and even in these instances, the final type has become apparent within usually days to months. In addition, any "evolution" is unidirectional (eg, from type 2 to type 3) and, under the classification scheme, would simply result in a reattempt of the same therapy (eg, embolization) or more invasive treatment (eg, surgery). Accordingly, the classification system assists in the planning and selection of initial and any future therapeutic interventions for all children, as it did in the present case.

Clinical Features

More than 30 pediatric cases of congenital IAPF involving the hepatic artery have been previously reported (Table 2), with its first report nearly 40 years ago (31). Overall, slightly more than half consisted of unilateral lesions. Three quarters presented by 2 years of age, with a mean age of approximately 3 years (range, 1 week to 16 years), which differs considerably from previous statements (26,32). Cases of unilateral IAPF presented at a mean of approximately 4 years, considerably later than cases of bilateral or complex IAPF.

Most symptoms and signs of IAPF are caused by the development of PH (5,17,33–37). At presentation, the most common symptoms are upper GI bleeding (66% of cases), failure to thrive (50%) and chronic diarrhea or steatorrhea (50%). The latter symptoms can precede GI bleeding (38), but PH predates the diagnosis. Variceal bleeding (2,39) has been observed to occur within a year of appearance of a fistula (40). At least 20% of cases manifest with protein-losing enteropathy with hypoalbuminemia and fat malabsorption.

Physical examination usually reveals signs of PH. Marked splenomegaly (63% of cases), hepatomegaly (41%), ascites or edema (47%), prominent abdominal distension (41%) and cutaneous collateral circulation or caput medusae are detected. Auscultation of a continuous murmur or bruit in the right upper quadrant (33,41,42) is present in at least half of cases. Rarely, high-output congestive heart failure (CHF) may occur in very young infants (1,31,37,43) because of left-to-right shunt through a patent ductus venosus (32), as flow is otherwise restricted by hepatic sinusoids interposed between the fistula and the right heart (1,4,44). CHF occurs more commonly in hepatic arteriovenous malformation (AVM), a communication between a systemic artery and a hepatic vein, as well as hepatic hemangiomata and hemangioendotheliomas (45,46), which are abnormal multiple connections between the hepatic artery and hepatic vein. As a result, infants with large hemangioendothelioma or hepatic AVMs tend to present much earlier, often in the neonatal period (47). Congenital IAPF differs from these lesions because it does not communicate directly with the systemic venous circulation.

Arterioportal malformations cannot usually be distinguished from more common hepatic lesions, such as hemangioma or hemangioendothelioma, without radiological studies (46). Differentiating between an IAPF and a systemic AVM is however vital because therapeutic decisions differ greatly for anatomical and physiological reasons.

The sustained macrocytosis in the present case was difficult to explain, with exclusion of obvious causes. We felt that mild hepatic dysfunction (48) and reticulocytosis (49) were likely responsible.

Pathophysiology

The variable presentation of congenital IAPF is related to the variations in fistula location and shunt size (1,2,43,50). Severity of symptoms is proportional to the volume of blood that is shunted (1,4). Chronic malabsorption, diarrhea and abdominal pain are attributed to mesenteric vascular congestion (31,32,35,51). Abnormal protein loss due to ischemic small bowel has also been demonstrated experimentally (52). In addition to protein-losing enteropathy (53), steatorrhea or evidence of fat malabsorption may occur and may

TABLE 2. *Clinical features of the congenital intrahepatic arterioportal fistula syndrome (N = 32) (1,2,5,31–38,41–43,50,51,53,55,57–60,62–64,66,73,74)*

Clinical Feature	
Gender—% cases (n)	
Male	66 (21)
Female	34 (11)
Type of IAPF—% cases (n)*	
Unilateral	53 (17)
Bilateral	22 (7)
Complex	19 (6)
Unreported	6 (2)
Mean age (range) at presentation‡	
Overall	35 mo (1 wk–16 y)
Unilateral	53.5 mo (3 wk–16 y)
Bilateral	12.5 mo (1 wk–6 y)
Complex	15.3 mo (3 mo–6 y)
Primary symptoms—% cases	
Upper GI bleeding	66
Failure to thrive	50
Chronic diarrhea/steatorrhea	50
Vomiting	22
Clinical signs—% cases	
Splenomegaly	63
RUQ bruit	47
Ascites or edema	47
Hepatomegaly	41
Abdominal distension	41
Laboratory investigations—% cases	
Anemia	69
Hypoalbuminemia	22
Occult blood-positive stool	19
Definitive intervention—% cases (n)	
Embolization(s) alone	47 (15)
Surgical alone	28 (9)
Embolization and surgical	19 (6)
Transplantation	3 (1)
Nil	3 (1)
Outcome—% cases (n)	
Well	88 (28)
Alive with PPH	9 (3)
Died	3 (1)

*Type: U, unilateral; B, bilateral; C, complex; see Table 1.
‡Age at which symptoms first appeared.
IAPF, intrahepatic arterioportal fistula; GI, gastrointestinal; RUQ, right upper quadrant of abdomen; PPH, persistent portal hypertension; y, years; mo, months; wk, week(s).

contribute to malnutrition. Fat malabsorption may be a manifestation of lymphatic vessel dilatation and leak (51) and possibly pancreatic hypofunction stemming from abnormal portal circulation (51,53). Most importantly, long-standing impaired portal flow can lead to portal vein thrombosis and subsequent cavernous transformation of the portal vein, and obliterative changes such as hepatoportal sclerosis and fibrosis of portal radicles can develop in the portal microvasculature (33,36,51).

As antegrade flow diminishes in the portal vein, hepatic artery flow increases, in part because of the hepatic arterial buffer response (51). Some authors (31,37,51) have suggested that excessive flow in the HA may contribute to hypoperfusion of organs distal to its origin, a "steal" phenomenon that compromises superior and inferior mesenteric circulation. Hypoperfusion may result in further small bowel edema and ischemia (37,50,51), hemorrhage (51) and, in some cases, small bowel infarction with necrosis (31). Dehydration and anemia are exacerbating factors (32). Subtotal villous atrophy has been noted in only one other case of IAPF (6). Intestinal biopsies in children with IAPF generally show some combination of vascular dilatation, interstitial edema and fibrosis of the lamina propria (32), but occasionally can be normal (35). Some findings can mimic those of intestinal lymphangiectasia (51). Most likely, the villous atrophy we identified was nonspecific (51) and attributable to malnutrition.

No genetic basis for this syndrome has been described thus far.

Imaging

Ultrasonography is a rapid, reliable and noninvasive modality for identifying prehepatic PH and is the first-line imaging modality in IAPF (3,20,32,33,37,51,54–56). Hepatic angiography serves to confirm the diagnosis and accurately delineate the vascular anatomy to allow for ablation of the fistula (1,26,35,50,51,57–59). Selective arteriography of the hepatic arteries is accomplished by digital subtraction and allows for planning of treatment, either surgical or embolization. In addition to the IAPF and its feeding vessels, pulsatile flow is observed in the portal vein and fistula, with reversal of flow during diastole.

Management

To date, no cases of congenital IAPF closing spontaneously have been reported. Definitive therapy is aimed at obliteration of the shunt and restoration of normal portal and hepatic arterial hemodynamics (32,60). The options for treatment are (1) surgical, (2) percutaneous transarterial embolization (TAE) (1,39,61) with various materials and (3) a combination of TAE and surgery. Surgical approaches have included ligature of the implicated hepatic arteries or branches (5,33,35,37,50), fistula excision and direct vascular repair (62) and partial hepatectomy (5,33,44,63). The latter is associated with higher mortality and morbidity (60,64).

Therapy selection depends on fistula location (33,36), lesion extent (2,41,51), accurate localization of all shunts (65,66), available materials and hospital experience with transcatheter embolization (2,33,35,65). Interventional radiological procedures now afford a relatively safe, inexpensive and practical method for treatment of IAPF (1,5,7,13,36,39,41,42,60,67,68). Compared with surgery, TAE carries the advantage of reduced morbidity, repeated access availability and reduced costs (1,2,36,60). Shorter hospital stay and decreased pain are specific advantages. Embolization is

technically possible if the arterial supply proximal to the IAPF can be accessed (1). In young infants, thrombosis of the femoral artery during or after angiograpy (41) is a significant risk of TAE; thus, systemic anticoagulation in this group is warranted. Risk of portal vein thrombosis may also be increased transiently after procedures that reduce portal venous flow. Accordingly, heparin was used at angiography in the present case. Care should also be taken to avoid compromising future surgical or transplant options.

All of the 10 published cases of unilateral lesions in which embolization was attempted were definitively treated by this method. TAE is very likely to be effective for unilateral lesions if only one feeding artery is present (2,41). Among the 7 patients with bilateral lesions, 3 were successfully treated by embolization alone, whereas 3 failed embolization. Surgery was definitive in the other 4 of 7 bilateral cases, 2 of which were treated definitively by ligation. Some authors (1,6,17) have posited that bilateral lesions should be treated by ligation of feeding arteries, with embolization affording only short-term palliation. However, repeated endovascular interventions are often necessary to be curative, particularly if multiple bilateral feeding arteries are present or if collaterals subsequently develop (2,19,33,66).

Complex congenital IAPF are prone to collateralization or recurrence after radiological intervention (32). Hence, they may be difficult to treat without a combination of surgery and embolization. Ours was the first case (1 of 4) of a complex IAPF to be eradicated by embolization attempts alone. Only half (2 of 4) of ligation attempts for complex IAPF have been successful. Two complex cases required partial hepatectomy after failing other treatments, and another underwent elective liver transplantation after failed embolization.

As shown in the present case, smaller branches with low flow can be effectively occluded with superselective injection of particulate or liquid embolic agents, such as particulate polyvinyl alcohol sponge (Ivalon) (36) or *N*-butyl 2-cyanoacrylate glue (66). Such materials can be combined with detachable metallic coils (66) and may be underused in IAPF. Detachable microcoils and balloons (60) alone are also suitable for smaller arterial branches. These should be positioned as close as possible to the fistula in the feeding artery (69,70). Distal positioning may minimize the risk of recanalization of the fistula by other collateral arteries and also preserve transarterial access to distal collateral arteries that are later identified (66).

Authors of some early reports felt that coil embolization of a large fistula carried a high risk of portal vein thrombosis (35). This could potentially occur if coils migrated into the portal system (2). However, portal vein thrombosis has not been observed as an adverse effect of embolization in treatment of congenital IAPF. Nonetheless, much care must be taken with large IAPF (diameter $\geq$4.5 mm) through which flow is high. A 6/4-mm Amplatzer occlusion device, also used to occlude patent ductus arteriosus, has been used judiciously in this setting (2). Injury to bile ducts due to repeated embolization, even of complex lesions, has not been reported.

Persistent symptoms in children for whom multiple transarterial embolizations have failed necessitate surgical intervention with ligation where possible or, potentially, partial hepatic resection. Portosystemic shunts, often used in treating systemic AVMs, should not be undertaken in the treatment of IAPF, as they may precipitate CHF by rerouting arterialized portal blood directly into the systemic venous circulation (71), with loss of protection conferred by hepatic sinusoids.

Routine follow-up US (72) is necessary to verify cessation of flow through the IAPF and stability of embolization material. Thereafter, the frequency of US monitoring remains to be established. The fistula may recur symptomatically within days to a few years after treatment (32,33,36,66), warranting ultrasonographic study. Often, this "recurrence" is actually an unmasking of other coexisting hepatoportal shunts whose reconstitution of the fistula becomes apparent only after ablating a dominant communication (73), as occurred in the present case. Angiography is usually repeated if symptoms recur within a few days of the attempted treatment (74).

Overall, TAE is the mainstay of treatment in most children with noncomplex congenital IAPF (1,14,15,20,36,44,58,60) and is curative alone in nearly half of such cases. A surgical approach should be considered for fistulae that do not resolve after several trials of embolization (14,33,50,64). Until sufficient comparative data are available, surgery should be considered as the initial treatment of patients with complex IAPF.

Prognosis

With the exception of the first published case of congenital IAPF, all patients have survived. Most children become asymptomatic shortly after treatment and show excellent catch-up growth, with decompression of the portal circulation. Significant morbidity occurs in some patients, especially with delays in diagnosis. Hepatic synthetic function is preserved in nearly all children with congenital IAPF (1,2,32,53).

Persistent portal venous hypertension with cavernous transformation is reported in less than 10% of published cases. All were infants 4 months of age or younger with complex (type 3) IAPF (33,36,63), suggesting complex morphology with early onset as a negative prognostic feature. This event occurred in all such infants despite multiple therapeutic endeavors, including TAE. In one instance (36), cavernous transformation with insidious thrombosis of the portal vein occurred 2 years after

embolization, but importantly, hepatofugal portal flow had persisted after symptomatically effective coil ablation. Prolonged exposure to systemic arterial pressures likely resulted in sinusoidal injury, PH and eventual portal vein thrombosis (36).

CONCLUSIONS

Congenital intrahepatic arterioportal fistula, a rare but treatable cause of PH, should be considered in an infant or child with recurrent and severe upper gastrointestinal bleeding, failure to thrive, hepatic bruit and splenomegaly or ascites. Recognition of the development of this unusual constellation of features will avoid a potentially costly delay in diagnosis and treatment. Irreversible PH is a consequence of long-standing IAPF. Ultrasonography with Doppler flow is a first-line investigation for this disorder. Treatment decisions may be based on the proposed classification system for this syndrome. Interventional radiological procedures are the preferred treatment for most children with noncomplex lesions, but multiple trials and a variety of embolization materials may be necessary to achieve a cure.

REFERENCES

1. Vauthey JN, Tomczak RJ, Helmberger T, et al. The arterioportal fistula syndrome: clinicopathologic features, diagnosis, and therapy. *Gastroenterology* 1997;113:1390.
2. Kumar N, de Goyet Jde V, Sharif K, et al. Congenital, solitary, large, intrahepatic arterioportal fistula in a child: management and review of the literature. *Pediatr Radiol* 2003;33:20–3.
3. Ryckman FC, Alonso MH. Causes and management of portal hypertension in the pediatric population. *Clin Liver Dis* 2001;5: 789–818.
4. Van Way CW III, Crane JM, Riddell DH, et al. Arteriovenous fistula in the portal circulation. *Surgery* 1971;70:876–90.
5. Heaton ND, Davenport M, Karani J, et al. Congenital hepatoportal arteriovenous fistula. *Surgery* 1995;117:170–4.
6. Heaton ND. Hepatic arterioportal fistulas: surgical ligation or embolization? [reply]. *Surgery* 1996;119:237.
7. Tanaka H, Iwai A, Sugimoto H, et al. Intrahepatic arterioportal fistula after blunt hepatic trauma: case reports. *J Trauma* 1991;31: 143–6.
8. Kayser S, Marincek B, Schlumpf R, et al. Rapidly progressive portal hypertension 23 years after post-traumatic arterioportal fistula of the liver. *Am J Gastroenterol* 1996;91:1442–6.
9. Sziklas JJ, Spencer RP. Hepatic artery-portal vein fistula detected on hepatic flow study: case report. *J Nucl Med* 1975;16:910–1.
10. Eastridge BJ, Minei JP. Intrahepatic arterioportal fistula after hepatic gunshot wound: a case report and review of the literature. *J Trauma* 1997;43:523–6.
11. English WP, Johnson MB, Borman KR, et al. Mesenteric ischemia: an unusual presentation of traumatic intrahepatic arterioportal fistula. *Am Surg* 2001;67:865–7.
12. Okuda K, Mucha H, Nakajima Y, et al. Frequency of intrahepatic arteriovenous fistulae as a sequela to percutaneous needle puncture of the liver. *Gastroenterology* 1978;74:1204–7.
13. Isik FF, Greenfield AJ, Guben J, et al. Iatrogenic arterioportal fistulae: diagnosis and management. *Ann Vasc Surg* 1989;3:52–5.
14. Lumsden AB, Allen RC, Sreeram S, et al. Hepatic arterioportal fistula. *Am Surg* 1993;59:722–6.
15. Pashankar D, Schreiber RA, Culham G. Arterioportal fistula in a child with biliary atresia. *Gastroenterology* 1998;114:862–3.
16. Stringer MD, McClean P, Arthur RJ, et al. Congenital hepatoportal arteriovenous fistula [letter]. *Surgery* 1996;118:235–6.
17. Stringer MD, McClean P, Heaton ND, et al. Congenital hepatic arterioportal fistula [letter]. *J Pediatr Gastroenterol Nutr* 1999; 29:487–8.
18. Mieles LA, Waldman D, Orloff MS. Arterioportal fistulae following segmental liver transplantation in a child. *Pediatr Transplant* 2002;6:419–22.
19. Foster JH, Sandblom P. Portal hypertension secondary to an hepato-portal arteriovenous fistula. *Ann Surg* 1961;154:300–4.
20. Tarazov PG, Prozorovskij KV. Intrahepatic spontaneous arterioportal fistula: duplex ultrasound diagnosis and angiographic treatment. *Am J Gastroenterol* 1991;86:775–8.
21. Onitsuka A, Katagiri Y, Shibata M, et al. Congenital multiple arterioportal fistula: a case report treated with portocaval shunt. *Hepatogastroenterology* 1997;44:1324–7.
22. Bolognesi M, Sacerdoti D, Bombonato G, et al. Arterioportal fistulas in patients with liver cirrhosis: usefulness of color Doppler US for screening. *Radiology* 2000;216:738–43.
23. Fontaine J, Groose A, Tongio J, et al. A propos d' un case de fistule arterio-veineuse hepatico-portale. *Chirurgie* 1970;96: 235–40.
24. Sachdeva R, Yapor M, Schwersenz A. Massive variceal bleeding caused by a hepatic artery–portal vein fistula: a manifestation of hepatocellular carcinoma in a 12-year-old. *J Pediatr Gastroenterol Nutr* 1993;16:468–71.
25. Kanazawa S, Niiya H, Mitogawa Y, et al. Cavernous hemangioma with arterioportal and arteriohepatic vein shunts coexisting with hepatocellular carcinoma. *Radiat Med* 1994;12:83–5.
26. Gallego C, Miralles M, Marin C, et al. Congenital hepatic shunts. *Radiographics* 2004;24:755–72.
27. Martini GA. The liver in hereditary haemorrhagic telangiectasia: an inborn error of vascular structure with multiple manifestations: a reappraisal. *Gut* 1978;19:531–7.
28. Roman CF, Cha SD, Incarvito J, et al. Transcatheter embolization of hepatic arteriovenous fistula in Osler-Weber-Rendu disease: a case report. *Angiology* 1987;6:484–8.
29. Zentler-Munroe PL, Howard ER, Karani J, et al. Variceal hemorrhage in hereditary haemorrhagic telangiectasia. *Gut* 1989; 30:1293–7.
30. Bodner G, Peer S, Karner M, et al. Nontumorous vascular malformations in the liver. *J Ultrasound Med* 2002;21:187–97.
31. Gryboski JD, Clemett A. Congenital hepatic artery aneurysm with superior mesenteric artery insufficiency: a steal syndrome. *Pediatrics* 1967;39:344–7.
32. Marchand V, Uflacker R, Baker SS, et al. Congenital hepatic arterioportal fistula in a 3-year-old child. *J Pediatr Gastroenterol Nutr* 1999;28:435–41.
33. Meunier C, Dabadie A, Darnault P, et al. Congenital intrahepatic arterio-portal fistula: diagnostic and therapeutic aspects. *Pediatrie* 1993;48:211–6.
34. Machado MCC, da Cunha JEM, Bacchella T, et al. Congenital intrahepatic arteriovenous fistula associated with portal hypertension and digestive bleeding. *Rev Hosp Clin Fac Med Sao Paulo* 1979;34:285–8.
35. Iñón AE, D'Agostino D. Portal hypertension secondary to congenital arterioportal fistula. *J Pediatr Gastroenterol Nutr* 1987;6:471–3.
36. Routh WD, Keller FS, Cain WS, et al. Transcatheter embolization of a high-flow congenital intrahepatic arterial-portal venous malformation in an infant. *J Pediatr Surg* 1992;27:511–4.
37. Bakker J, Robben SGF, Hazebroek FWJ, et al. Congenital arterioportal fistula of the liver with reversal of flow in the superior mesenteric vein. *Pediatr Radiol* 1994;24:198–9.
38. D'Agostino D, Mónaco RG, Alonso V, Iñón A, Ciardullo M, de Santibañes E. Liver transplantation as treatment for arterioportal fistulae. *J Pediatr Surg* 1998;33:938–40.

39. Tarazov PG. Intrahepatic arterioportal fistulae: role of transcatheter embolization. *Cardiovasc Intervent Radiol* 1993;16:368–73.
40. Foley WJ, Turcotte JG, Hoskins PA, Brant R, Ause R. Intrahepatic arteriovenous fistulae between the hepatic artery and portal vein. *Ann Surg* 1971;174:849–55.
41. Fiane AE, Gjestvang FT, Smevik B. Hepatoportal arteriovenous fistula and bleeding oesophageal varices in a child. *Eur J Surg* 1993;159:185–6.
42. Lamireau T, Chateil J-F, Petit P, et al. Successful embolization of congenital intrahepatic arterioportal fistula in two infants. *J Pediatr Gastroenterol Nutr* 1999;29:211–4.
43. Cheung YF, Leung MP. Coil embolization of hepatoportal arteriovenous fistula in a neonate. *Clin Cardiol* 1999;22:675–6.
44. Fellows KE, Hoffer FA, Markowitz RI, et al. Multiple collaterals to hepatic infantile hemangioendotheliomas and arteriovenous malformations: effect on embolization. *Radiology* 1991;181:813–8.
45. Howard ER, Heaton ND. Haemangiomas. In: Howard ER ed. *Surgery of Liver Disease in Children*, 1st ed. Oxford, UK: Butterworth-Heinemann, 1991:115–25.
46. Boon LM, Burrows PE, Paltiel HJ, et al. Hepatic vascular anomalies in infancy: a twenty-seven-year experience. *J Pediatr* 1996;129:346–54.
47. Shumacker HB, Waldhausen JA. Intrahepatic arteriovenous fistulae of hepatic artery and portal vein. *Surg Gynecol Obstet* 1961;112:497–501.
48. Mates M, Heyd J, Souroujon M, et al. The haematologist as watchdog of community health by full blood count. *QJM* 1995;88:333–9.
49. Pappo AS, Fields BW, Buchanan GR. Etiology of red blood cell macrocytosis during childhood: impact of new diseases and therapies. *Pediatrics* 1992;89:1063–7.
50. Helikson MA, Shapiro DL, Seashore JH. Hepatoportal arteriovenous fistula and portal hypertension in an infant. *Pediatrics* 1977;60:921–4.
51. D'Agostino D, Orsi M. Congenital hepatic arterioportal fistula [letter]. *J Pediatr Gastroenterol Nutr* 1999;29:487.
52. Zuidema GD, Turcotte JG, Wolfman EF Jr, et al. Metabolic studies in acute small-bowel ischemia. *Arch Surg* 1962;85:130.
53. Agarwala S, Dutta H, Bhatnagar V, et al. Congenital hepatoportal arteriovenous fistula: report of one case. *Surg Today* 2000;30: 268–71.
54. Gallego C, Velasco M, Marcuello P, et al. Congenital and acquired anomalies of the portal venous system. *Radiographics* 2002;22:141–59.
55. Altuntaş B, Erden A, Karakurt C, et al. Severe portal hypertension due to congenital hepatoportal arteriovenous fistula associated with intrahepatic portal vein aneurysm. *J Clin Ultrasound* 1998;26:357–60.
56. Xia JG, Dong SX, Li FH, et al. Color Doppler ultrasonic diagnosis of congenital portal vein malformations. *World Chinese Journal of Digestology* 2004;12:1376–9.
57. Lin SL, Lee CM, Kong MS, et al. Congenital hepatic arterioportal fistula complicated with gastrointestinal bleeding treated with transcatheter embolization: case report. *Chang Gung Med J* 1999;22:106–10.
58. Raghuram L, Korah IP, Jaya V, et al. Coil embolization of a solitary congenital intrahepatic hepatoportal fistula. *Abdom Imaging* 2001;26:194–6.
59. Alkim C, Şahin T, Oğuz P, et al. A case of congenital intrahepatic arterioportal fistula. *Am J Gastroenterol* 1999;94:523–25.
60. Akpek S, Ilgit ET, Cekirge S, et al. High-flow arterioportal fistula: treatment with detachable balloon occlusion. *Abdom Imaging* 2001;26:277–80.
61. Applbaum YN, Renner JW. Steel coil embolization of hepatoportal fistulae. *Cardiovasc Intervent Radiol* 1987;10:75–9.
62. Gorenflo M, Waldschmidt J, Bein G, et al. Arterioportal fistula in infancy. *J Pediatr Gastroenterol Nutr* 1993;16:87–9.
63. Fasching G, Schimpl G, Sauer H, et al. Necrotizing enterocolitis due to congenital arterioportal fistulas in an infant. *Pediatr Surg Int* 1993;8:264–7.
64. Billing JS, Jamieson NV. Hepatic arterioportal fistula: a curable cause of portal hypertension in infancy. *HPB Surgery* 1997;10: 311–4.
65. Tarazov PG. Hepatic arterioportal fistulas: surgical ligation or embolization? *Surgery* 1996;119:237.
66. Cil BE. Transhepatic embolization of a recanalized congenital hepatic arterioportal fistula with NBCA and coils. *Cardiovasc Intervent Radiol* 2004;27:172–4.
67. Nakamura T, Isobe Y, Ueno E, et al. Successful arterial embolization of arteriovenous fistula in the portal circulation. *Gastrointest Radiol* 1992;17:324–6.
68. Desai SR, Khan S, Thomas S, et al. *The 4 year review of hepatic angiography at a centre for paediatric liver disease and transplantation II*. Vienna, Austria: Eighth European Congress of Radiology, 1993:317.
69. Redmond PL, Kumpe DA. Embolization of an intrahepatic arterioportal fistula: case report and review of the literature. *Cardiovasc Intervent Radiol* 1988;11:274–7.
70. Romeo JM, Herrera N, Picouto LP. Angiographic embolization of intrahepatic arterioportal fistula. *Br J Surg* 1991;78:736–7.
71. Ramchandani P, Goldenberg NJ, Soulen RL, et al. Isobutyl-2-cyanoacrylate embolization of a hepatoportal fistula. *AJR Am J Roentgenol* 1983;140:137–40.
72. Taourel P, Perney P, Bouvier Y. Angiographic embolization of intrahepatic arterioportal fistula. *Eur Radiol* 1996;6:510–3.
73. Protheroe S, Tanner MS, Bolia A. Ultrasound demonstration and transhepatic embolization of congenital aortoportal fistula with hepatoportal arteriovenous malformation in an infant. *J Interv Radiol* 1993;7:157–62.
74. Kang TC, Lee HC, Yang FS, et al. Idiopathic hepatic arterioportal fistula: report of one case. *Acta Paediatr Taiwan* 2002; 43:102–5.