Arterioportal Fistulas: Introduction of a Novel Classification With Therapeutic Implications

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Arterioportal fistulas (APFs) are arteriovenous communications between the splanchnic arteries and the portal vein that represent an infrequent cause of presinusoidal portal hypertension. They can be acquired or congenital. Penetrating hepatic trauma, including liver biopsies, represent the most common etiology. They can be asymptomatic or manifest with portal hypertension. An abdominal bruit is a valuable physical finding. Persistence of an APF can cause hepatoportal sclerosis and possibly portal fibrosis. A detailed radiologic evaluation is mandatory. One must differentiate between small peripheral intrahepatic APFs (type 1) and large central APFs (type 2). The former usually resolve spontaneously, whereas the latter can cause portal hypertension and hepatic parenchymal changes. Type 1 APFs caused by needle injury can be followed by Doppler ultrasound. All other fistulas need treatment. Arterioportal fistulas are first treated by transcatheter embolization. Surgical approaches are reserved for complex cases. Congenital APFs (type 3) are diffuse and intrahepatic and can be difficult to manage. Overall, the prognosis is good. Herein, we propose a novel classification for arterioportal fistulas with therapeutic implications. (J GASTROINTEST SURG 2006;10:543–550)

KEY WORDS: Arterioportal fistula, hepatoportal fistula, arteriovenous fistula, portal hypertension, embolization

Arterioportal fistulas (APFs) are a complex group of arteriovenous fistulas that have intrigued surgeons for over a century. The term encompasses all fistulas between any of the splanchnic arteries and the portal veins. They can either be acquired or congenital and can present with a myriad of clinical manifestations owing to their specific physical characteristics and flow pressure parameters. Although rare, the incidence of APF is rising secondary to increased interventional procedures to the liver[1–3] and the improved survival of patients with hepatic trauma.[4] The purpose of this report is to provide a comprehensive review of the literature and to introduce a new classification, with emphasis on therapeutic implications.

HISTORY

The first description of APF in the literature was by Weigert in 1886. In 1892, Sachs reported on a patient who died of bleeding esophageal varices and who was found to have a ruptured hepatic artery aneurysm on postmortem examination.[5–7] APF as a complication of liver biopsy was first described by Preger in 1967.[8] In 1971, Van Way et al.[6] comprehensively reviewed the clinical characteristics of these interesting arteriovenous fistulas. In 1997, Vauthey provided a contemporary analysis of the APF syndrome and made important therapeutic recommendations.[9]

CLASSIFICATION

APFs are classified by their etiology, size, involved vessels, and location. They can either be congenital or acquired, large or small, intrahepatic or extrahepatic, central or peripheral, traumatic or spontaneous. Their inflow can be from any of the splanchnic arteries, most commonly, the hepatic artery (hepatoportal fistula) (65%), followed in incidence by the splenic artery (splenoportal fistula) (11%) and the superior mesenteric artery (10%).[9] It is critical to classify the APF properly in order to select appropriate therapy. In this article we

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propose a new classification of APFs into types 1, 2, and 3 (Table 1).

Type 1

These are small, peripheral, intrahepatic fistulas with minimal physiologic consequences. The most common etiology is percutaneous liver biopsies. Patients are usually asymptomatic and their APF generally thromboses within 1 month. The suggested treatment is close follow-up with Doppler ultrasound. If the fistula persists for longer than 1 month or if the patient becomes symptomatic, transcatheter embolization is recommended.

Type 2

Larger, more central fistulas with enough flow to cause elevated portal pressures are type 2. They can be either intrahepatic or extrahepatic. Examples are those caused after penetrating abdominal trauma or by erosion of a splenic artery aneurysm into the portal system. They cause portal hypertension and hepatoportal sclerosis and can progress to portal fibrosis. These fistulas should be treated to avoid the complications of portal hypertension and prevent irreversible hepatic parenchymal changes. The suggested treatment is embolization if technically possible. If this is unfeasible or unsuccessful, then surgical approaches are warranted.

Type 3

These are congenital APFs. These rare fistulas are usually intrahepatic and diffuse, and they cause severe portal hypertension in infancy. We recommend referral to a specialized pediatric hepatobiliary center where treatment may consist of hepatic artery ligation, embolization, resection, or liver transplantation.

Table 1. Proposed classification of arterioportal fistulas (APFs)

	Definition	Example	Clinical Findings	Natural History	Treatment
Type 1	Small peripheral asymptomatic APF with minimal physiologic insult	APF caused by percutaneous liver biopsy	Asymptomatic	Thrombose spontaneously within 1 month	Follow up with Doppler ultrasound
					Embolize if persistence beyond 1 month or symptomatic
Type 2	Larger central fistulas causing physiologic insult	Delayed presentation of APF after penetrating abdominal trauma	Portal hypertension	Portal hypertension	Treat
			History of penetrating abdominal trauma	Hepatoportal sclerosis	Embolize if possible
			Abdominal bruit	Possibly portal fibrosis	Surgery if not feasible or unsuccessful
Type 3	Congenital fistulas	Diffuse, intrahepatic congenital APF in a neonate	Failure to thrive	Severe portal hypertension in infancy	Referral to specialized pediatric hepatobiliary center
			Portal hypertension		
			Diarrhea		