Meso-Rex Bypass—A Procedure to Cure Prehepatic Portal Hypertension: The Insight and the Inside

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Prehepatic portal hypertension (PHPH), related to thrombosis and the cavernomatous transformation of the portal vein (PVC), is the single most common cause of portal hypertension in children. Although it can be secondary to the direct damage that relates to neonatal catheterization of the umbilical vein, the latter condition represents <25% of cases,[1-7] and no cause is found in most cases (idiopathic PHPH). Even less commonly, it relates to regional trauma (or surgery, eg, after liver transplantation), tumors, or infection (eg, peritonitis, abscess). Although in the adult age group there is a clear correlation between pre-existing thrombophilia and PVC, coagulation abnormalities are not found to be primarily related to PVC in children, in fact, only minor disorder types have been reported in a small proportion of patients, and most coagulation profile abnormalities observed are acquired and secondary.[8-10]

Anatomically speaking, the initial thrombotic process seems to mainly involve the portal vein trunk, to which it is eventually limited in the typical disease condition; a variable extension, either downstream into the intrahepatic radicals, or upstream into the splanchnic system, or both, can be observed.[1,4,11-13] Remarkably, in children who have had catheterization of the umbilical vein, the processes more than likely start in an inverse manner, with the thrombus initiating within the liver as a consequence of the direct (chemical, physical, or infection) damage of the intrahepatic veins and, more precisely, the left portal system.[2,7,12] A diffuse splanchnic venous thrombosis is exceptional in children (although it is relatively common in adults with thrombophilia). Although part of the portal system is not reachable for decompressive surgery (as the portal trunk and sometimes other veins are thrombosed), various types of portosystemic shunt have been performed with success; the type of shunt depends on the venous anatomy of each patient and the team's preferences (mesocaval and various splenorenal types of shunt).[14-20]

Disclosure Information: Nothing to disclose.

Received September 15, 2013; Revised October 30, 2013; Accepted October 30, 2013.

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The meso-Rex bypass (MRB), also called the mesentericoportal bypass, is the most recent procedure introduced in the surgical armamentarium for managing such conditions. Although MRB was initially proposed (in 1992) for curing thrombosis of the portal vein after pediatric liver transplantation,[21-25] its indication rapidly extended to cure idiopathic PHPH in children who had a cavernoma due to PVC in the presence of a healthy liver.[26-35] Briefly, the operation consists of bypassing the thrombosed portal trunk (and the cavernoma) by interposing a graft between the superior mesenteric vein (SMV) and the recessus of Rex (Figs. 1 and 2); the latter recessus is, anatomically speaking, a part of the left portal system, the precise portion that runs sagittally within the umbilical scissure between segments II, III, and IV (Couinaud classification). The latter recessus is the portion of the portal system that was, before birth, part of the umbilical venous pathway (formed by the umbilical vein, the Rex recessus, and the Arantius channel) (Fig. 2).[21,36] Because of its natural position and the related precise landmarks (umbilical ligament and umbilical scissure), it is a segment of the intrahepatic portal system that is both relatively easy to access surgically and away from the cavernoma (which typically develops within the porta hepatis).

After its introduction 20 years ago, the technique gained interest and many teams around the world started implementing this original approach into their clinical practice; good outcomes have been reported repeatedly, the major advantage of the procedure being a natural and physiological cure (by restoring the hepatopetal portal flow[29]) of portal hypertension, contrary to conventional portosystemic shunts that are palliative procedures, as they treat portal hypertension by diverting the splanchnic blood flow away from the hepatic route. The MRB reduces portal pressure by reopening an access to the low-resistance hepatic parenchyma. In turn, MRB allows the regression of natural portosystemic connections and reverses the side effects, if any, related to either portal hypertension and/or portosystemic connections[13,32-35] (Table 1). It has been shown to reverse hepatopulmonary syndrome,[30] improve neurocognitive ability,[37] help normalize hyperammonemia,[33] or reverse encephalopathy caused by portosystemic connections.[38] Meso-Rex bypass can prevent formation of liver nodules and adenoma in children with natural or surgical portosystemic shunts,[4,20]

ISSN 1072-7515/13/$36.00
http://dx.doi.org/10.1016/j.jamcollsurg.2013.10.024

Abbreviations and Acronyms

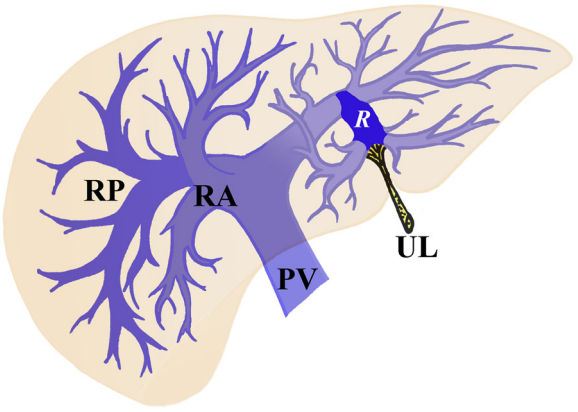
MRB = meso-Rex bypass
PHPH = prehepatic portal hypertension
PVC = cavernomatous transformation of the portal vein
SMV = superior mesenteric vein

and reverse commonly observed coagulopathy in patients with cavernoma (elevated prothrombin time and international normalized ratio), with decreased levels of factors V, VII, protein C, and protein S.[13] Lastly, it improves somatic growth in those patients who have displayed growth retardation.[39,40]

The MRB was recently recommended as the first-choice surgery for children with extrahepatic portal hypertension, a healthy liver, and a patent Rex recessus.[1,4,14,41] Some authors propose that this technique be performed earlier in the course of PHPH as preemptive management, thereby preventing many of the PHPH comorbidities or complications that develop with time and age.[1,4,32-34,37]

Although the procedure was first described 2 decades ago[22,23] and has been referred to or mentioned in many articles, this surgery has remained the prerogative of a relatively small panel of hepatobiliary or transplantation surgeons. In addition, the procedure has never been described in great detail and it has not been included in any textbook of surgical techniques. Based on our experience, we would like to describe the key points of the procedure for successful surgery. To the best of our knowledge, this is the first step-by-step detailed description of the standard MRB technique.

Figure 1. Retrograde portogram (image on the upper left): conventional mesenteric venogram, in the same patient, showing that the former imaging delineates a patent intrahepatic system, where the latter modality was not evidenced by the intrahepatic portal system. The meso-Rex bypass procedure restores a hepatopetal (physiological) route for the splanchnic venous blood through the reconnection of the 2 venous systems (arrows).



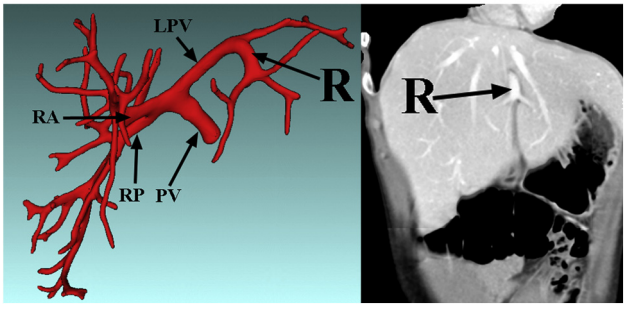


Figure 2. The recessus of Rex is that short (3-cm length) portion of the left portal vein running sagittally within the umbilical scissure between the left lateral segment (or segment II and III according to Couinaud) and the left anterior segment (segment IV). The anterior part of the recessus of Rex (R) ends in the umbilical remnant and ligament. LPV, left portal vein; PV, portal vein; RA, right anterior portal vein; RP, right posterior portal vein; UL, umbilical ligament.

METHODS

Preoperative assessment

All PHPH children are considered potential candidates for MRB and, therefore, all are assessed using a standardized diagnostic clinical, laboratory, and radiologic protocol, as follows.

Although it is rarely associated in children, thrombophilia must be excluded by a detailed study of the coagulation, including measurement of prothrombin time; partial thromboplastin time; international normalized ratio; anti-thrombin III; factors II, V, VII, and X; protein C and S; and a search for mutations of factor V Leiden; factor II prothrombin; and methylene-tetra-hydro-folate-reductase.[8-10,13]