# VARIATIONS OF THE REX BYPASS FOR EXTRAHEPATIC PORTAL VEIN OBSTRUCTION. REVIEW OF THE LITERATURE

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**Abstract**

*Introduction.* Portal vein thrombosis is the main cause of Portal Hypertension among children. Its ethology is heterogeneous and not completely understood and many cases of portal vein thrombosis are called idiopathic. With the introduction of Meso-Rex bypass 27 years ago, the outcome changed drastically as this shunt surgery restore portal blood to the liver. Since then, more and more surgeons use it to treat Portal Hypertension and they report variants of the original shunt operation in an effort to develop the best approach. *Objective.* This review paper aim to present the rex bypass and its variants reported so far, highlighting the best and the worst outcome. *Methods.* We have reviewed the English literature for articles presenting Extrahepatic Portal Vein Obstruction treated with Rex shunt surgery. Articles were reviewed systematically. For limitation of bias we have excluded Case Report articles and Review articles in which the authors or their affiliation institution have published more than one article using same patient population or collected data from articles used in this study.

**Keywords:** Rex shunt, extrahepatic portal vein obstruction, graft, variant Rex shunt

## Introduction

Portal vein thrombosis (PVT) is the main cause of portal hypertension (PHT) in pediatric population [1]. The term PVT refers to obstruction (complete or incomplete) of the portal venous flow due to an intraluminal thrombus. When the obstruction is limited to extrahepatic segment of the porta hepatica is referred to as Extrahepatic Portal Vein Obstruction (EHPVO), although many authors use PVT and EHPVO terms interchangeably [2]. Incriminated factors that lead to thrombosis of the portal vein are numerous (see Table 1) [3] and the treatment is constantly evolving as the underlying disease is better studied and understood. Patients with PVT without a intrinsic liver disease (e.g. cirrhosis) have a normal liver function but display growth retardation, coagulopathy, alteration of neurocognitive function and other symptoms related with liver deprivation of normal portal venous flow. As collateral circulation develops rapidly, bleeding form esophageal and gastric varices may be the first symptom of PHT which can be fatal. The role of shunt surgery is well established in the treatment of PHT and among many shunt alternatives the Rex shunt created in 1992 by Jean de Ville de Goyet and his team became the preferred option for selected patients as it reestablish portal flow to the liver with the potential of cure. Although Rex bypass is not clearly stated in the treatment guidelines is it highly indicated for selected patients as the best option for shunt surgery [2].

Table 1. Etiological factors encountered in PVT in pediatric population.

| Etiological factor |
|---|
| Portal vein injury<br>Umbilical vein catheterization<br>Trauma, splenectomy, pancreatic surgery, colectomy, etc |
| Post liver transplantation |
| Local inflammatory conditions<br>Pancreatitis<br>Abdominal sepsis<br>Liver abscesses |
| Coagulation disorders<br>Factor V Leiden mutations (rs6025)<br>Prothrombin gene mutation (G20201A)<br>MTHFR gene mutation (C677T)<br>Hyperhomocysteinemia<br>Protein C deficiency<br>Protein S deficiency<br>Antithrombin III deficiency<br>Antiphospholipid syndrome/Anticardiolipin antibodies |
| Post biliary atresia operation (portoenterostomy) |
| Idiopathic |

## Objective

At the end of this review, the reader will be familiar with physiologic and pathologic anatomy of the portal vein system as in Extrahepatic Portal Vein Obstruction.

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Will know the Rex shunt and its variations available in the literature for performing this type of shunt surgery which is, currently, the preferred surgical treatment option for selected patients. We aim to highlight the Rex Shunt techniques with best results in short and long-term outcome.

**Methods**

We have reviewed the English literature for articles presenting Extrahepatic Portal Vein Obstruction treated with Rex shunt surgery. For finding relevant articles we have searched for "Rex Shunt", "Rex Bypass", "Meso – Rex" and "Mesenterico Left" and the results – more than 100 articles were reviewed systematically. For limitation of bias we have excluded Case Report articles and Review articles in which the authors or their affiliation institution have published more than one article using same patient population or collected data from articles used in this study. While reviewing the relevant articles we have focused on the operative technique, the type of graft used as a conduit and short and long-term outcome of the operation highlighting complications.

*Normal and pathologic anatomy.*

In brief, during the 4th to 6th embryonic life the omphalomesenteric veins transport blood form the gut and the umbilical veins transport blood form the placenta to the embryo. Blood is transported through the hepatic sinusoids in the liver, then through the hepatic veins in the heart. Some blood bypass the liver through the ductus venosus. The left omphalomesenteric vein will become the portal vein as the right one will involute by the end of 6th week. Also the right umbilical vein will disappear and the left umbilical vein will be patent until after birth when it thromboses and become round ligament of the liver. Portal vein forms at the confluence of the superior mesenteric and splenic veins just posterior to the head of the pancreas. In the splenic vein drains the Inferior mesenteric vein anywhere along its course. In the portal vein drains the coronary (left gastric) vein which communicates with distal esophageal veins (will become esophageal varices in PHT). Portal vein divides into the right and left portal branches. In the right portal branch drains the cystic vein (which will contribute to cavernous transformation of the portal vein). The portal vein will collect blood from the intestines, stomach, pancreas, spleen and gall bladder [4]. The normal blood flow through the portal vein is approximately 18% of systemic blood flow [5]. When the thrombosis occur, the liver blood supply is diminished by up to half, but the hepatic arterial buffer rapidly compensate perfusion and so, the acute thrombosis can take place without displaying any symptoms especially if there is an acute disease at this time (e.g. sepsis) which masks the symptoms, if any, of acute PVT. Collateral circulation to bypass obstruction rapidly develop, usually within a 3-5 weeks period. Most important collateral pathways are depicted below:

(1) left gastric (coronary) vein and short gastric veins to esophageal veins and thenceforth to azygous and hemiazygous veins,

(2) superior hemorrhoidal veins to the middle and inferior hemorrhoidal veins into the inferior cava vein (IVC),

(3) umbilical vein to epigastric veins trough superficial veins of the abdominal wall,

(4) in the retroperitoneum, intestinal veins to branches of IVC,

(5) veins (Sappey) around the falciform ligament to epigastric or intrathoracic veins.

These collaterals dilate and become varicose. Anyhow, of most interest are the esophageal and gastric varices because these can rupture due to lumen dilatation, increased wall tension, thin wall and ulceration. Acute EHPVO is considered to be if the symptoms appear within the first 2 months form the thrombosis event in the absence of PHT or Portal Cavernoma (PC). Chronic EHPVO is when there is a PHT (with or without PC) or its complications like variceal bleeding, hypersplenism, ascites. Cavernous transformation of the Portal vein is a sequel of acute thrombosis and usually, but not always, define chronicity. PCs are reported as early as few days form the thrombosis and there are also patients without a PC even after 2 months. For better understanding of this heterogeneous pathological transformations at the Sixth Baveno Consensus (April 2015) for PHT, experts presented a classification for portal vein obstruction (see Table 2).

Table 2. Classification of portal vein obstruction.

| | |
|---|---|
| Site of PVT | Type 1: Only trunk<br>Type 2: Only branch: 2a- One, 2b – both<br>Type 3: Trunk and branches |
| Presentation | R: Recent<br>Ch: Chronic |
| Underlying liver disease | C: Cirrhotic<br>N: Non-Cirrhotic liver disease<br>H: HCC and other local malignancies<br>L: Post liver transplant<br>A: Absence of underlying liver disease |
| Degree of portal venous system occlusion | I: Incomplete<br>T: Total/ Complete |
| Extent of PV occlusion | Splenic vein, Mesenteric vein, Both |

Using this classification could help deciding on treatment, diagnostic modalities, prognostication and asses the outcome of shunt surgery on short and long term [2].

Classic Meso-Rex bypass. Mesenterico-to-left-portal vein bypass was initially designed to cure portal vein thrombosis after liver transplantation in children, but soon after it begun to be used with success for EHPVO in children with a healthy liver. In current practice, all children with PVT are candidates for a Rex bypass. Careful and complete preoperative assessment must be performed. Must be excluded intrinsic liver disease (e.g. cirrhosis) and thrombophilia. Also, the Rex recessus must be patent otherwise the shunt will not be possible to perform. The Rex recessus is that short part of the left portal vein located within the umbilical scissure (between hepatic segments II, III, IV) in a sagittal orientation. If you follow the round ligament into the liver, one will find and open the Rex recessus as its anterior part is related to the ligament. In brief, the operation as designed by de Ville de Goyet is as follow: usually the liver is addressed through a bilateral subcostal incision, followed by its luxation anteriorly and medially. Round and falciform ligaments are divided to access the Rex recessus. A portion of the liver parenchyma is resected on each side of the umbilical scissure with great attention not to damage portal branches of the II and III segments. Dissecting the round ligament of the liver in the umbilical sccisure will reveal the rex recessus. Preparation of the recessus is performed close to the vein wall until all the collateral branches and the origin of left portal vein are identified. At this point an angiography can be performed to confirm patency of the Rex recessus. After the preparation of Rex recessus, the superior mesenteric vein and the route for the bypass conduct preparation is performed. Next step is to harvest internal jugular vein to be used as a conduit. The Rex recessus and its collaterals are clamped with suture ties and a Satinsky clamp and a longitudinal venotomy is made. The subclavian end of the graft is anastomosed to the Rex recessus in an end to side manner followed by removal of the clamps. The bypass is completed with the end to side anastomosis at the superior mesenteric part. This is type 1 of Meso-Rex bypass. De Ville de Goyet also described a type 2 bypass when he use right gastroepiploic vein to anastomose it in the Rex recessus without the need of a harvested graft [1,6]. For very detailed and step-by-step description of the operation we suggest reading "Meso-Rex Bypass – A procedure to Cure Prehepatic Portal Hyprtension: The Insight and the Inside" by di Francesco, Grimaldi and de Ville de Goyet.

We analysed data and operative details on a total of 494 patients operated using different Rex shunt techniques (see Table 3 [5,7–25]) and the most used technique (59%) is the "classic" Jean de Ville de Goyet's Meso-Rex bypass using internal jugular vein as a conduit. It seems to give the best outcome on short and long term follow-up. Most frequent complications reported are the shunt thrombosis and stenosis. While some thrombosis events were resolved by thrombolysis, especially if it was an early event, some were permanent and required portosystemic shunt surgery. Stenosis of the shunt was usually corrected with percutaneous dilatation or stent placement. Overall shunt patency approaches 90% in the literature. Most complications were diagnosed by close follow up. Some clinical aspects that suggested a possible complication was recurrence of variceal bleeding episodes, persistent or recurrent splenomegaly and poor weight gain.

*Variant Rex Shunt surgery*

Recent literature articles present experiences with variants of the original Meso-Rex bypass. Most notable results are reviewed in this paper.

Transposition of gastric coronary vein, splenic vein, recanalized umblilical vein or inferior mesenteric vein to complete the bypass without the need to harvest a graft was observed in 18% of studied cases. In the largest study, Zhang et al [8,26] performed a gastro-portal bypass on 48 cases. They mobilized the coronary vein and anastomosed it to the left portal vein. When suitable (diameter of the coronary vein > 0, 5 cm) they concluded is the preferred method. This type of Rex bypass has some significant advantages, like less vascular anastomoses, less scaring and preservation of the jugular veins. The results are comparable with classic meso-rex bypass, except for the variant when recanalized umblilical vein is used, as this has a higher complication rate. Although the preliminary results are very good, more and detailed data is necessary to conclude.

More of an ad-on to the Rex bypass than a variant of the bypass, is the paraeophageal and paracardial devascularisation to alleviate esophageal and gastric varices. Wang et al propose that, at the time of the bypass, first step is to perform excision of paraesophagogastric veins. This include coronary vein, short gastric veins, posterior gastric vein, left inferior phrenic veins and ectopic high esophageal branches. They also recommend splenectomy or partial splenectomy if the patient is over 6 years of age and in the presence of hypersplenism. They concluded that this procedure is effective for preventing variceal bleeding and portal gastropathy. Also they consider it reduces the risk of bypass thrombosis because of increased portal flux and pressure [18].

In some cases the jugular vein is not available or is not long enough. Alternative grafts that can be used include, cadaveric cryopreserved iliac vein, great saphenous vein, splenic vein, inferior mesenteric vein, recanalized umbilical vein, gastroepiploic vein, jejunal or ileal vein, polytetrafluoroethylene (PTFE or Gore-Tex®) synthetic grafts and coronary vein. It appears to be of great significance the type of graft used, as in one series, Krebs-Schmitt et al reported that all cryopreserved iliac veins and umbilical vein got thrombosed at a median time of 21 months (rangeing between one day and 69 months). Even after recanalization of the shunt, these got rethrombosed. They concluded that best outcome is observed when jugular vein is used [19]. Regarding the use of saphenous vein, Louto et al presented a thnique where the graft consists of both great saphenous veins longitudinally cuted and sutured together around a Hegar to create a tubular graft of large diameter after the valves were excised. The outcome reported is that 7 out of 21 cases developed thrombosis. One

on 8th postoperative day and the remaining 6 developed late thrombosis (median range 20 months). Their thrombosis rate seems to be above 30% compared to the 10% rate when using jugular vein. Theoretical factors that predispose to thrombosis are the many cuts in the graft (longitudinal cuts and valve excision cuts) [16]. The information avaliable about the use of synthetic grafts like PTFE are sparse and insufficient. In our reviewed papers we only encountered 3 cases in which was used PTFE without mentioning any complications.

Table 3. No. – number of Rex shunts studied/performed by the author.

| *Author / year* | *No.* | *Type of graft used to complete the bypass* | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | JV | ePTFE | Al. | Sph. V | SV | IMV | GV | UV | None | NS / Others |
| *Timothy B Lautz/ 2013* | 70 | 70 | | | | | | | | | |
| *Ruo-Yi Wang/2017* | 42 | 10 | | | 8 | 2 | | 2 | | 20 | |
| *D. Krebs-Schmitt/2009* | 25 | 17 | | 5 | | | | | 3 | | |
| *Wei Chen/2011* | 5 | 5 | | | | | | | | | |
| *Daniel A. Bambini/2000* | 5 | 4 | | 1 | | | | | | | |
| *Nelson E. M. Gibelli /2011* | 11 | 11 | | | | | | | | | |
| *Mark D. Stringer/ 2007* | 11 | 11 | | | | | | | | | |
| *Tae-Yong Ha/2015* | 12 | | | | | | | | | 12 | |
| *Caroline Rochon/2012* | 6 | | | | | | | | | 6 | |
| *Jin-Shan Zhang/2017* | 79 | | | | | | | | | 53 | 26 |
| *Sukru Emre/2009* | 3 | 2 | 1 | | | | | | | | |
| *Li Long/2017* | 5 | | | | | | 4 | | | | 1 |
| *Gloria Chocarro/2016* | 18 | 16 | | 2 | | | | | | | |
| *Rukhmi Bhat/2013* | 65 | 56 | | | | | | | | | 9 |
| *Zhang Wei/2014* | 22 | | | | | 22 | | | | | |
| *Paloma Triana Junco/2018* | 19 | 19 | | | | | | | | | |
| *Jorg Fuchs/2003* | 7 | 7 | | | | | | | | | |
| *Topi Luoto/2012* | 21 | | | | 21 | | | | | | |
| *F. Gúerin/2013* | 43 | 40 | 2 | | | | | 1 | | | |
| *Heather A. Stefek/2018* | 25 | 22 | | | | | 3 | | | | |
| *TOTAL* | 494<br>100 % | 290<br>59 % | 3<br><1% | 8<br>1,6 % | 29<br>6% | 24<br>5% | 7<br>1,4% | 3<br><1 % | 3<br><1% | 91<br>18% | 36<br>7% |

JV – jugular vein internal or external (autograft); ePTFE – expanded polytetrafluoroethylene / Gore-Tex® (synthetic graft); Al. – allograft (cryopreserved iliac vein); Sph.V. – saphenous vein; SV – splenic vein; IMV – Inferior mesenteric vein; GV – gastroepiploic vein or coronary vein; UV – umbilical vein; None – no graft was used to complete the shunt, but the transposition of specified vessels; NS/ Others – not specified or other type of graft.

**Conclusion**

Overall conclusion is that classic meso-rex bypass is the best option so is indicated whenever possible. The outcome compared to the other variant rex bypasses is better and the diameter of the graft might have a role which is yet to be demonstrated. The diameter of the graft influences the blood velocity and at least theoretical, a higher velocity could reduce the thrombosis risk. Further research on determining the optimal diameter of the graft in order to properly release portal hypertension and yet to maintain a high velocity of the blood in the graft need to be conducted. At last, synthetic graft like PTFE needs to be studied as they may play an important role as they are available on-demand, but using a synthetic graft that will not grow or expand over time when a child grows might limit its use to the adult patient.

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