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When to consider surgery in the treatment of portal hypertension

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Historical aspects of portal hypertension

- The early experience with portosystemic shunts led to the following practice patterns and beliefs:
 - 1. Surgery should only be considered as a last resort
 - 2. Surgery should only be considered after a **bleeding episode**
 - **3. Splenectomy** is reasonable treatment for hypersplenism due to portal hypertension
 - Repeated endoscopic procedures ± general anesthesia is preferable to major surgery
 - Any surgery should be delayed until a child is <u>older (bigger)</u> because the results are better in bigger patients



History of surgery for portal hypertension

- Banti in Florence first linked portal hypertension to hypersplenism
- Eck (1847 1908) in Germany developed concept of porto-caval fistula



Arch Fr Pediatr. 1955;12(6):590-602.

Am J Surg. 1964 Mar;107:447-51.

PORTAL DECOMPRESSION PROCEDURES IN CHILDREN.

CLATWORTHY HW Jr, DELORIMER AA.

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Classification of portal hypertension



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- 1. Extra (pre) hepatic
 - a) Idiopathic
 - b) Cavernous transformation
 - c) AV fistulae
- 2. Sinusoidal
 - a) Liver disease
 - b) Hypoplastic portal vein (Klatzkin's Disease- hepatopedal sclerosis)
- 3. Outflow obstruction
 - a) Budd Chiari



Causes of portal hypertension in children

- Primary biliary or portal plate malformations
 - Biliary atresia
 - Congenital hepatic fibrosis
 - PFIC
- Hepatocellular
 - Cystic Fibrosis
 - Alpha-1 antitrypsin deficiency,
 - NASH
 - Veno-occlusive disease
- Primarily extrahepatic disorders
 - Extrahepatic portal vein obstruction (EHPVO)
 - Budd-Chiari syndrome
 - Congestive heart failure (e.g., Fontan related)



Parenchymal Disease

- 1. Cystic Fibrosis
- 2. Congenital Hepatic Fibrosis in the setting

of ARPKD and possible future kidney

transplant

3. Biliary Atresia

- Non selective shunts
 - Divert most of the mesenteric venous return into the systemic circulation
 - Porta caval shunt
 - Mesocaval shunt
 - Proximal splenorenal shunt
 - TIPS
 - Serious side effects possible in patients with intrinsic liver disease
- Selective
 - Distal splenorenal shunt



Splenorenal shunt

- Selective shunt
 - Warren and Zeppa 1967
 - Created a shunt without stealing from the portal supply to the liver
 - Less mortality
 - Less encephalopathy







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Parenchymal Disease- CF

· · · Engl

			Cystic Fibrosis			
	Variables		Pre DSRS	Post DSRS	p value	arenchyma
			(n=5)	(n=5)		
	Pulmonary function tests	Mean FEV1 (% predicted) (sd)	67.6 ± 25.7	56.0 ± 27.6	0.068	nsion
	Portal hypertension	Mean Spleen size (cm) (sd)	8.4 ± 1.52	4.4 ± 1.82	0.019*	:ransplant
		Mean Platelet count (10E9) (sd)	88 ± 48	92 ± 35	0.9	
	Growth	Mean z-scores Height (sd)	0.30 ± 2.31	1.12 ± 1.97	0.15	ated by
		Mean z-scores Weight (sd)	-0.30 ± 1.91	0.35 ± 1.39	0.17	
		Mean z-scores BMI (sd)	-0.18 ± 1.11	-0.04 ± 0.08	0.36	lunting
	Liver function tests	Mean INR (sd)	1.29 ± 0.13	1.21 ± 0.08	0.36	
		Mean Albumin (g/dL) (sd)	3.58 ± 0.37	3.28 ± 0.33	0.24	[]
		Mean Total Bilirubin (mg/dL) (sd)	1.46 ± 0.44	1.94 ± 1.31	0.53	

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Distal Splenorenal Shunt in CF patients





Congenital Hepatic Fibrosis



- Portal Hypertension secondary to parenchymal disease
- Often accompanied by advanced renal disease that will require kidney transplant
- Logic: leukopenia and thrombocytopenia not a good setting for a kidney transplant
- Distal splenorenal shunt for reversal of hypersplenism rather than liver transplantation
- Liver function usually well preserved.



Treatment of pre-kidney transplant portal hypertension





53 patients shunted for PH due to parenchymal disease





Follow up for DSRS



- Post op day 1 US
- Every 3 months
 - US
 - CBC platelet count
 - Physical exam to follow spleen size
- 1 year CT scan
- Yearly physical exam
- Important negative

No routine follow up endoscopies.



What is the outcome after DRSR for portal hypertension in patients with advanced fibrosis/cirrhosis?

• Biliary atresia

- Transplanted 19, 6 and 2^{*} years after DSRS

	Total	Doing well	Trx/died
Biliary atresia	10	7	3
CHF	24	10	2/2
CF	9	8	0/1
OTHERS	10	8	2/0

CHF

- 2 transplants 5 years (liver and kidney) and 1 year after DSRS
- 1 death after a renal transplant of unknown causes with patent shunt, and 1 death after combined liver-kidney transplant

• CF

 1 death of progressive lung disease with patent shunt and stable liver function

* Hepatopulmonary sydrome



EHPVT

- Congenital or acquired obstruction of the extra-hepatic portal vein
 - Cavernomatous transformation of the portal vein
 - Portal vein thrombosis
- Not an acute disorder (no anticoagulation indicated)
- Associated with umbilical vein catheterization
- omphalitis
- 25 % incidence of congenital heart
- Associated with hypercoagulable condition
- May affect both extra hepatic mesenteric veins, as well as, intrahepatic portal vein branches





- The portal vein: important for liver wellness: 75% of hepatic blood flow (20 % of Portal hypertension Porto systemic shunting 50 % of oxygen to the liver
- Bleeding from varies to trophic factors to sustain normal false liver growth and function neurotransmitters
 - Esophageal
 - Gastric

- What happens when the portaReptown of the so-active is under high pressure or deficient ances from the portal
- Intestinal
- Rectal
- Hypersplenism
 - Enlarged spleen
 - Thrombocytopenia
 - Pain from splenic capsule stretching
 - Limitation of activities

 Removal of circulating immune complexes.

(ammonia)

circulation

 Normal somatic growth suffers



Mesenteric to Left Portal vein bypass



- Described in 1996 as treatment for portal vein thrombosis after whole liver transplant
- Vein graft from superior mesenteric vein to the intrahepatic left portal vein
- Later extended to patients with non transplant related PVT



Pre operative assessment



Preoperative imaging Ultrasound and MR or CT venogram Retrograde transjugular venogram Liver function tests and liver biopsy Coagulation profile PT, platelets, coagulation factors, hypercoagulable states Factor V, VII, VIII, AT III, protein C and S, plasminogen Leiden mutation, antiphospholipid antibodies Neurodevelopmental testing Cardiac echocardiogram

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Retrograde transjugular portal venogram



- 1. Determine quality of the intrahepatic portal vein
- 2. Measure hepatic, free and wedge pressures
- 3. Obtain a liver biopsy





Who to operate on: experience countsdealing with the poor intrahepatic portal vein





Changes in intrahepatic portal vein over time





Pre and post op CT scan





Marginal inflow and marginal intrahepatic PV





Marginal inflow and marginal intrahepatic PV cont'd





Marginal inflow and marginal intrahepatic PV – cont'd



- Baveno B- D venograms do not always predict failure
- Exploring the Rex recessus is always the best way of evaluating the possibilities
- Inflows may vary:
 - Inferior mesenteric vein
 - Large varices in the lesser sac
 - Other large intra-abdominal collateral;s may be very good inflow vessels.

Normal liver



Congenital hepatic fibrosis



EHPVT



hypertension





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Pre and post comparison



2 Argentina 1 Australia 4 Brazil 15 Canada 1 Chile 1 Cuba 2 Germany 1 India 2 Israel 1 Mexico 1 Pakistan 1 Puerto Rico 1 Ukraine 1 Vietnam



Patients with Idiopathic EHPVT

	Primary- open	Converted - open	Thrombosed	Total
Rex	143	4	2	149
Splenorenal	38	7	1	46
Mesocaval	7	11	0	18
Sugiura	1	2	0	3
Total	189	24	3	216

Overall Summary

Patients with EHPVT (Idiopathic, Post-transplant and Other)

	Primary- open	Converted - open	Thrombosed	Transplanted	Total
Rex	158	4	4	2	168
Splenorenal	42	11	1	1	55
Mesocaval	7	11	0	0	18
Sugiura	1	2	0	0	3
Total	208	28	5	3	244

Regression analysis between platelet count and portal flow as a function of cardiac output

Portal vein flow as a function of cardiac output

Outcome: GI bleeding Idiopathic portal vein thrombosis (161)

	Meso Rex n=115	PSS n= 43	Sugiura n= 3
# with preoperative bleeding	69	25	3
Recurrence of bleeding	1	0	1

- 2/161 patients with recurrent bleeding.
- Meso Rex and PSS equally effective at controlling bleeding complications from esophageal and gastric varices

Outcome: Hypersplenism

Outcome: Liver function

Correlation between age and shunt flow

r=-0.223 p=0.039

Overall summary

Procedures done for patients with EHPVT (n=209)

190 idiopathic EHPVT

■ open ■ DSRS ■ MC ■ Sugiura ■ throm bosed

Sugiura procedure: Arch Surg 112:1317, 1977

- Gastric devascularization
- Esophageal transection
- Splenic preservation in children
- Vagal sparing
- Partial wrap
 - Superina RA et al: A modified Sugiura operation for bleeding varices in children. Journal of pediatric surgery 1983;18(6):794-9.

TIPSS

transjugular intrahepatic porto systemic shunt

- Temporizing shunt
- Bridge to transplant
- Not a good long term solution in children
 - Hepatic decompensation
 - Shunt thrombosis
 - encephalopathy

Mesocaval shunt

- Indications
- Same as for the DSRS except when there is no suitable splenic vein
 - Portal vein thrombosis with no suitable intrahepatic portal vein
- We use ringed Gore-Tex grafts that are resistant to pressure and compression by surrounding tissues

Mesocaval shunt with artificial grafts

Surgical Options

Prophylaxic shunt for EHPVO

- 1. Patient qualifications for MRB include favorable anatomy and medical condition defined as
 - 1. "A" type anatomy confirmed by wedge two-dimensional portography (retrograde internal jugular venogram) by an experienced and multidisciplinary team (Fig. <u>3</u>)
 - Normal hepatic venous pressure gradient (HVPG) determined at the time of wedged portography and/or confirmatory liver biopsy revealing no fibrosis or underlying liver disease that would impact portal flow
 - 1. Patent superior mesenteric vein and splenic vein without thrombosis confirmed using sitespecific expertise in either computed tomographic or magnetic resonance angiography
 - 2. Bilateral internal jugular vein patency with equivalent symmetry confirmed by imaging such as ultrasound or computed tomographic angiography
 - 3. Hematological evaluation excluding an intrinsic prothrombotic condition that would not be reversed by MRB
 - 4. Body weight >8 kg
 - 5. Echocardiography excluding significant cardiovascular abnormalities and significant pulmonary hypertension
- 2. Surgical/institutional qualifications for performing preemptive MRB include
 - 1. Pediatric liver transplant and vascular and hepatobiliary expertise with multidisciplinary care including pediatric hepatology and interventional radiology
 - 2. Prior successful MRB experience

Conclusions

- 1. The surgical treatment of children with portal hypertension depends on the underlying state of the liver
- 2. The Rex shunt or mesenteric to left portal vein bypass is preferable to any portosystemic shunts for treatment of portal vein thrombosis
- Distal splenorenal and mesocaval shunting provides acceptable palliation in children with EHPVO and children with other forms of non cirrhotic and cirrhotic portal hypertension with good long term results
- 4. The Rex shunt provides
 - Relief from life threatening symptoms of portal hypertension
 - Correction of physiological functions of the liver including restoration of coagulation factor synthesis and reversal of encephalopathy

Baveno Bambino

<u>http://www.baveno7.com</u>

