Portal hypertension: the Children’s Memorial Experience

Portal hypertension: the pressure in the mesenteric venous bed exceeds 12 mm Hg.

- Bleeding: associated with increased mortality
- Hyper-splenism:
  - Limits physical activity
  - Bleeding from low platelets
  - Infections from leukopenia
  - Pain from splenic capsule
- Encephalopathy:
  - Behavioral issues
  - Learning disabilities in children
  - Sleep pattern alteration

Secondary Symptoms

- Ascites
- Porto-pulmonary syndrome
  - Persistent hypoxemia
- Pulmonary hypertension
  - Pulmonary vasoconstriction

History

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

- Central shunts
  - Porto-caval shunts
    - Eck fistula (end to side) - complete portal diversion from liver
    - Side to side - portal blood diverted from liver in proportion to the degree of portal hypertension
  - H graft (Sarfeh shunt)
    - Variant of the meso-caval, partial diversion of mesenteric blood
    - Central splenorenal + splenectomy

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

Classification of portal hypertension

- Extra (pre) hepatic
  - Idiopathic
  - Cavernous transformation

- Sinusoidal
  - Increased fibrous tissue in sinusoids leads to increased resistance to both inflow and outflow

- Outflow obstruction
  - Budd Chiari
  - Venous occlusive disease

- Increased inflow
  - Arterio venous fistula between artery and portal circulation

Emergency Treatment

- Volume replacement
  - Red cells, fresh frozen plasma, large bore lines

- Monitoring
  - ICU, CVP, Arterial lines

- Diagnosis
  - Endoscopy, arteriography, CT scanning

- Stop the Bleeding
  - Pharmacology
  - Local control
  - Pressure
  - Emergency surgery
Emergency Medical Treatment

- Octreotide (somatostatin)
  - As effective as balloon tamponade
  - Reduces hepatic blood flow, wedged hepatic vein pressure
  - Constrict splanchnic arterioles
- Blakemore tube
  - Direct pressure on esophageal and gastric varices

Emergency Medical Therapy

- Sclerotherapy
  - Injection of varices (number of solutions)
  - 91% control of acute bleeding
  - Repeated sessions necessary
  - High incidence of complications
- Banding
  - Safer and as effective as sclerotherapy

Long term treatment

- Beta blockers (propranolol, nadolol)
  - Reduces splanchnic blood flow
  - Reduces hepatic vein wedge pressure
- Nitrate vasodilators
  - isosorbide-5-mononitrate
- Effective in reducing repeat bleeding by more than 51%
- Most studies based on adult results

Prophylactic treatment

- Studies in both adults and children have demonstrated that prophylactic treatment of varices is justified
- Reduction in frequency of bleeding by either pharmacologic means or endoscopic ligation of varices once they have been diagnosed rather than to treat only once bleeding has already happened
- Surgery for prevention of initial bleed is more controversial if not done with another primary aim such as treatment of severe hypersplenism.
Surgical Shunts for treatment of portal hypertension: depends on severity of disease

- Advanced liver disease
  - Liver transplantation
- Stable liver disease/cirrhosis
  - Distal splenorenal shunt
- Pre hepatic portal obstruction
  - Rex shunt
    - Restores physiological homeostasis
- Post hepatic obstruction
  - Porta-caval/mesocaval
  - Liver transplant

TIPS (transjugular intrahepatic portosystemic)

- Relieves portal hypertension from intrinsic liver disease
- Shunt clots in time
- Reserved for children with advanced liver disease while waiting for liver transplant

Devascularization procedure: Sugiura

- Reserved for patients that cannot be shunted
- Interrupts para and intra gastroesophageal varices
- Spleen preserved in children
- Few side effects

Arteriovenous fistula

- Can cause torrential bleeding because of very high portal venous pressure
- Selective embolization of arterio-venous fistula
- Surgical ligation
Distal splenorenal shunt

- Congenital hepatic fibrosis - 5
- Portal vein thrombosis - 4 (1 child w/ Hep C)
- Hepatitis B - 1
- Wilson's disease - 1

11 children portal hypertension
Prerequisites for spleno renal shunt

- Patent splenic vein
- Patent renal vein
- CTA or MRV has replaced conventional angiography to demonstrate vascular anatomy

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

MR or CT angiography

- non invasive
- Imaging of intrahepatic portal vein more reliable in low flow states
- measure liver volume
- global assessment of intra-abdominal vasculature

Minimal requirements for successful shunt:
1. Preserved intrahepatic PV
2. Adequate vein for inflow
Patient characteristics

- 59 patients referred over 7 year period
- None excluded based on unsuitable anatomy defined by imaging

Distribution of patients

Patient characteristics

- 41 patients
  - 28 Bleeding
  - 12 hypersplenism
    » Profound thrombocytopenia
    » Recurrent infections
    » Activity restriction
  - 1 Encephalopathy

- Average age at diagnosis 4.1±3.3 (2 months - 13 year)
- Average age at surgery 7.9±4.9 (3 months - 19 years)

1. Dissect segmental branches of vein
2. Complete anastomosis of vein graft
3. Completes SMV anastomosis
Post op regimen

- Heparin until eating
- Aspirin and dipyridamole prophylaxis 6 months
- US during first week
- MRV prior to discharge
- Reassess 3, 6, 9, 12 months with bloodwork and imaging

Outcome according to cause of portal vein thrombosis

- 31/34 idiopathic open (92%)
  - 2 converted to Warren shunt
  - 1 no possible alternate shunt
- 1/3 Post transplant (33%)
  - 2 required retransplantation (both split grafts originally)
- 1/3 Other (33%)
  - 1 no possible alternate shunt
  - 1 converted to Warren

Correlation between age and restoration of mesenteric flow

\[ Y = 1.223 - 0.06 \times X \]

\[ R^2 = 0.317 \]

\[ p < 0.005 \]
Spleen size changes after Rex shunt

<table>
<thead>
<tr>
<th>Mean</th>
<th>S.D.</th>
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<tbody>
<tr>
<td>pre Rex</td>
<td>11</td>
</tr>
<tr>
<td>1 year</td>
<td>3</td>
</tr>
<tr>
<td>2 years</td>
<td>4</td>
</tr>
</tbody>
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*p<0.0001 between pre Rex and 1 year paired t test

Changes in PV size after restoration of mesenteric flow

- Rapid expansion of portal vascular bed
- Increase in size of intrahepatic portal vein branches
- Progressive increase in flow through the Rex shunt

Left portal vein diameter increases progressively after shunting by ultrasound measurement

<table>
<thead>
<tr>
<th>Mean</th>
<th>Std. Dev.</th>
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<tbody>
<tr>
<td>time 0</td>
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<tr>
<td>1 week</td>
<td>3.8</td>
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<tr>
<td>6 months</td>
<td>5.4</td>
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<tr>
<td>1 year</td>
<td>6.1</td>
</tr>
<tr>
<td>year 2</td>
<td>7.4</td>
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</table>
1. The surgical treatment of children with portal hypertension depends on the underlying state of the liver.

2. The Rex shunt is preferable to any portosystemic shunts for treatment of portal vein thrombosis.

Conclusion:
- The surgical treatment of children with portal hypertension depends on the underlying state of the liver.
- The Rex shunt is preferable to any portosystemic shunts for treatment of portal vein thrombosis.
Conclusions continued

1. Warren shunt is a good alternative in children with liver disease or children with unsuitable anatomy
2. Surgery should be considered early in the course of the disease as the success of shunting is high and the complication rates low