PORTAL HYPERTENSION

Sean Rim
SUNY Downstate
11/9/07
Case Presentation

- xx year old male with massive hematemesis presents to ED 8AM
- PMH
  - HTN
  - Hep B and C
  - EtOH abuse
- Admitted xx months ago for similar episode
- GCS 15 but BP 60/30 and P 70
- Minimal response to aggressive IVF resuscitation
- Exam significant for blood in airway, jaundice, ascites
- Intubated, started on pressors and octreotide
INR 1.8
AST 135
Surgery and GI consulted
Emergent EGD performed revealing multiple bleeding varices in mid and distal esophagus
Band ligation performed successfully
• Bleeding resumes after several hours
• Sengstaken-Blakemore tube placed with hemostasis
Patient brought to VIR for attempted TIPS
- Failed to show patent portal vein
Patient brought to MICU vented, on pressors
- 12 units PRBC
- 10 units FFP
- 3 units PLT

HCT remained stable

ABG 7.18/39.2/395/14.3/99%/-12.2
Despite continued aggressive resuscitation, patient expired on day xx secondary to multiorgan failure
“In cases of jaundice it is a bad sign when the liver becomes hard”

-Hippocrates

4th century B.C.
Cirrhosis

- Toxins
- Viruses
- Prolonged cholestasis
- Autoimmune
- Metabolic
- Hepatocellular necrosis
  - Fibrosis
    - Nodular regeneration
- Variceal hemorrhage
- Encephalopathy
- Ascites
- Hypersplenism
Autopsy studies reveal incidence between 3.5% and 5%.

10% to 15% of heavy drinkers develop alcoholic cirrhosis.

Sixth leading cause of death in 4th to 6th decades.
● Hepatic blood flow 1500 ml/minute

● 25% cardiac output

● Portal pressure
  ○ Hypertension >5 mm Hg
  ○ Portosystemic collateralization 8 to 10 mm Hg
- 1/3 of hepatic blood flow
- >1/2 of oxygen supply
- Direct regulation
- Catecholamines
- Maintains near constant total hepatic blood flow
- 2/3 of total hepatic blood flow
- Indirect regulation
- Constriction and dilation of splanchnics
Prehepatic portal hypertension
  - Portal vein thrombosis
  - Cavernomatous transformation

Splenic vein thrombosis
  - Gastrosplenic venous hypertension
  - Gastric varices secondary to gastroepiploic
Intrahepatic portal hypertension

- Schistosomiasis (presinusoidal)

- Alcoholic (sinusoidal and postsinusoidal)
  - Deposition of collagen in the space of Disse
  - Nodular regeneration distorts small hepatic veins

- Budd-Chiari, constrictive pericarditis, CHF (postsinusoidal)
Diagnostic goals

- Determine cause
- Estimate hepatic functional reserve
- Define portal venous anatomy
- Identify the site of GI hemorrhage
History

- Alcohol abuse
- Hepatitis
- Exposure to hepatotoxins
● History
  ○ Alcohol abuse
  ○ Hepatitis
  ○ Exposure to hepatotoxins

● Physical
  ○ Jaundice
  ○ Chest wall spider angiomas
  ○ Ascites
  ○ Caput medusae
  ○ Asterixis
Laboratory studies

- Pancytopenia (hypersplenism)
- Anemia
- Electrolyte abnormalities
- Coagulopathy
- Elevated LFT’s
Prognosis of variceal bleeding
- Noncirrhotic 5% to 10%
- Cirrhotic 40% to 70%
- 30% rebleed within 6 weeks
- 70% rebleed within 1 year
<table>
<thead>
<tr>
<th>Clinical and Laboratory Measurement</th>
<th>Patient Score for Increasing Abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1</td>
</tr>
<tr>
<td>Encephalopathy (grade)</td>
<td>None</td>
</tr>
<tr>
<td>Ascites</td>
<td>None</td>
</tr>
<tr>
<td>Bilirubin (mg/dL)</td>
<td>1–2</td>
</tr>
<tr>
<td>Albumin (g/dL)</td>
<td>≥3.5</td>
</tr>
<tr>
<td>Prothrombin time (increase, sec)</td>
<td>1–4</td>
</tr>
</tbody>
</table>

Grade A, 5-6; Grade B, 7-9; Grade C, 10-15
<table>
<thead>
<tr>
<th>Child Class</th>
<th>Mortality</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>5%</td>
<td>28%</td>
</tr>
<tr>
<td>B</td>
<td>25%</td>
<td>48%</td>
</tr>
<tr>
<td>C</td>
<td>&gt;50%</td>
<td>68%</td>
</tr>
</tbody>
</table>
● MELD score

● Marker of disease severity in alcoholic cirrhosis and predictor of mortality

- $3.78 \ln{\text{serum bilirubin (mg/dL)}} + 11.2 \ln{\text{INR}}$
- $+ 9.57 \ln{\text{serum creatinine (mg/dL)}} + 6.43$

- Scored from 6 to 40
Patient has acute variceal hemorrhage

- Protect airway.
- Initiate resuscitation.
- Obtain I.V. access.
- Administer prophylactic antibiotics.

Give octreotide (250 µg bolus, then 25–50 µg/hr for 2–4 days).
Alternatively, give vasopressin (20 U bolus, then 0.2–0.4 µg/min) plus nitroglycerin (initially 50 µg/min, titrated to BP).

Hemorrhage is not controlled
- Perform endoscopic variceal ligation or endoscopic sclerotherapy.

Hemorrhage is controlled

Hemorrhage is not controlled
- Consider TIPS.

Hemorrhage is controlled
Patient has acute variceal hemorrhage

Protect airway.
Initiate resuscitation.
Obtain I.V. access.
Administer prophylactic antibiotics.

Give octreotide (250 µg bolus, then 25–50 µg/hr for 2–4 days).
Alternatively, give vasopressin (20 U bolus, then 0.2–0.4 µg/min) plus nitroglycerin (initially 50 µg/min, titrated to BP).

Hemorrhage is not controlled
Perform endoscopic variceal ligation or endoscopic sclerotherapy.

Hemorrhage is controlled

Hemorrhage is not controlled
Consider TIPS.

Hemorrhage is controlled
TIPS is available and successful

TIPS is unavailable or unsuccessful
Perform balloon tamponade.

Hemorrhage is not controlled
Assess patient's candidacy for transplantation.

Patient is future transplant candidate
Construct mesocaval interposition shunt or DSRS.

Patient is not future transplant candidate

Ascites is intractable
Construct side-to-side portosystemic shunt.

Ascites is absent or manageable
Construct end-to-side portacaval shunt.
• Gastric balloon 50 ml
- Gastric balloon 50 ml
- AXR
- Gastric balloon 50 ml
- AXR
- 300 ml
- Gastric balloon 50 ml
- AXR
- 300 ml
- External traction
- Gastric balloon 50 ml
- AXR
- 300 ml
- External traction
- Esophageal balloon to 40 mm Hg
● Successful in 90% of cases
● 50% recurrence rates
● Treatment related mortality 20%
  ○ Perforation
  ○ Aspiration
  ○ Airway obstruction
Transjugular Intrahepatic Portosystemic Shunt

- Nonselective side-to-side portosystemic shunt
- Significant improvement in treating acute hemorrhage
- Cannulate hepatic vein via IJ
- Pass guidewire over needle through parenchyma into portal vein
Transjugular Intrahepatic Portosystemic Shunt

- Overall mortality not improved with TIPS alone
- Increased hepatic encephalopathy
- Contraindicated in right heart failure
Transjugular Intrahepatic Portosystemic Shunt

- Major limitation is shunt stenosis
  - 50% within the first year
  - 10% to 15% are total occlusions
- Some patients may not live long enough to experience stenosis
- “Bridge” to orthotopic liver transplants
Pomier et al. **TIPS versus endoscopic variceal ligation in the prevention of variceal rebleeding in patients with cirrhosis: a randomized trial.** Gut 2001; 48: 390-396

- Eighty cirrhotic patients with variceal bleeding (Child score 7-12)
- Randomly selected for TIPS or ligation after initial control of hemorrhage
● Ligation group
  ○ Days 1 and 10 then every 3 to 4 weeks
  ○ Followed every 3 months

● TIPS group
  ○ Performed within 72 hours
  ○ Followed monthly for 3 months then every 3 months
### Table 1  Patients characteristics at randomisation

<table>
<thead>
<tr>
<th></th>
<th>Ligation (n=39)</th>
<th>TIPS (n=41)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (y)</td>
<td>54.3 (10.9)</td>
<td>52.9 (13.3)</td>
</tr>
<tr>
<td>Sex (M/F)</td>
<td>27/12</td>
<td>29/12</td>
</tr>
<tr>
<td>Aetiology of liver disease (n)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alcoholism</td>
<td>24</td>
<td>25</td>
</tr>
<tr>
<td>Hepatitis B virus</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Hepatitis C virus</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>PBC</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Sclerosing cholangitis</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Autoimmune hepatitis</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Haemochromatosis</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Cryptogenic</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Previous variceal bleed (n)</td>
<td>8</td>
<td>10</td>
</tr>
<tr>
<td>Previous sclerotherapy (n)</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>Associated fundal varices (n)</td>
<td>6</td>
<td>8</td>
</tr>
<tr>
<td>Blood units (index bleed)</td>
<td>3.1 (2.5)</td>
<td>3.4 (2.9)</td>
</tr>
<tr>
<td>Time from index bleed to randomisation (hours)</td>
<td>42 (33)</td>
<td>44 (35)</td>
</tr>
<tr>
<td>Time from randomisation to TIPS (hours)</td>
<td>13 (11)</td>
<td></td>
</tr>
<tr>
<td>Balloon tamponade</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>Pugh score</td>
<td>9.8 (1.6)</td>
<td>9.6 (1.6)</td>
</tr>
<tr>
<td>Serum bilirubin (μmol/l)</td>
<td>89 (93)</td>
<td>49 (37)*</td>
</tr>
<tr>
<td>Serum albumin (g/l)</td>
<td>25.0 (5.3)</td>
<td>24.9 (4.5)</td>
</tr>
<tr>
<td>Haemoglobin (g/l)</td>
<td>91 (17)</td>
<td>89 (17)</td>
</tr>
<tr>
<td>Prothrombin (INR)</td>
<td>1.41 (0.28)</td>
<td>1.38 (0.25)</td>
</tr>
</tbody>
</table>

PBC, primary biliary cirrhosis; INR, international normalised ratio.

*p<0.05.
Rebleeding rates

18.5% versus 66%
Encephalopathy and Survival

47% versus 44%  
56% versus 53%
Surgical therapy

- Reserved for refractory cases in acute bleeding
- Esophageal transection with EEA
  - Avoid vagus nerve
  - Operative mortality and complications, 76% and 26%
● Portosystemic shunt
  ○ Wide variability in reported survival rates 30% to 70%
  ○ Manipulation of porta hepatis can complicate potential transplant surgery
Surgical therapy

- Most effective method of preventing recurrent variceal hemorrhage
  - Portosystemic shunts
  - Esophagogastric devascularization
  - Orthotopic liver transplant
Portosystemic Shunts

● Nonselective
  ○ End-to-side “Eck fistula”
  ○ Side-to-side
  ○ Interposition mesocaval shunt
  ○ Proximal splenorenal shunt

● Selective
  ○ Distal splenorenal shunt
End-to-side

- Only nonselective shunt thoroughly studied
- 9% to 25% rebleed versus 65% to 98%
- Trend towards increased survival
Side-to-side

- Decompresses sinusoids
- Improve ascites
Interposition Mesocaval Shunt

- Avoids hilar dissection
- Potential shunt ligation in cases of refractory encephalopathy
- Shunt thrombosis 35%
Proximal Splenorenal Shunt

- Splenectomy
- Proximal splenic vein into left renal vein
- Eventual hepatofugal flow into renal vein
Distal Splenorenal Shunt

- Anastamose distal splenic vein to left renal
- Interrupt venous collaterals
- Decompress esophagogastric circuit
- Mesenteric circuit provide hepatopedal flow
Distal Splenorenal Shunt

- Does not address mesenteric and sinusoidal hypertension
- Contributes to ascites formation
- Contraindicated in patients with
  - Refractory ascites
  - Splenic vein thrombosis
  - Prior splenectomy
  - <7 mm splenic vein diameter
Distal Splenorenal Shunt

- 50% of patients lose selectivity by one year
  - “Pancreatic siphon” effect
  - Splenopancreatic disconnection not yet adequately studied
  - Varied results in studies of encephalopathy and overall survival
Esophagogastric Devascularization

• Most effective nonshunt operation to prevent recurrence of variceal bleed
  ○ Esophageal transection
  ○ Splenectomy
  ○ Ligation of venous branches entering distal esophagus and proximal stomach
  ○ Selective vagotomy and pyloroplasty
Esophagogastric Devascularization

- Left gastric vein and paraesophageal collaterals preserved
- Permits portoazygous collateralization to inhibit future varix formation
<table>
<thead>
<tr>
<th></th>
<th>Japan</th>
<th>U.S.</th>
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</thead>
<tbody>
<tr>
<td>Operative Mortality</td>
<td>5%</td>
<td>20%</td>
</tr>
<tr>
<td>Rebleeding</td>
<td>6%</td>
<td>55%</td>
</tr>
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</table>
Orthotopic Liver Transplantation

- Most definitive therapy for complications of portal hypertension
- Child class A or mild B do relatively well with nontransplantation therapy
- Severe Child class B or C benefit more from early transplant

- Retrospective review of 1000 patients over 50 years
- Underwent surgical intervention for bleeding portal hypertension
Table 1. Types of Operation by Period

<table>
<thead>
<tr>
<th></th>
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<th></th>
<th></th>
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<tbody>
<tr>
<td>Selective shunts</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal splenoportal</td>
<td>0</td>
<td>114</td>
<td>83</td>
<td>56</td>
<td></td>
<td>253</td>
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<tr>
<td>Splenocaval</td>
<td>0</td>
<td>35</td>
<td>6</td>
<td>2</td>
<td></td>
<td>43</td>
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<tr>
<td>Low-diameter</td>
<td>0</td>
<td>0</td>
<td>29</td>
<td>10</td>
<td></td>
<td>39</td>
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<tr>
<td>Portacaval</td>
<td>96</td>
<td>5</td>
<td>0</td>
<td>2</td>
<td></td>
<td>103</td>
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<tr>
<td>Arterialized</td>
<td>0</td>
<td>10</td>
<td>0</td>
<td>0</td>
<td></td>
<td>10</td>
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<tr>
<td>Proximal splenoportal</td>
<td>35</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<td>35</td>
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<tr>
<td>Mesosystemic shunts</td>
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<td></td>
<td></td>
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<tr>
<td>Renal</td>
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<td>10</td>
<td>0</td>
<td>0</td>
<td></td>
<td>10</td>
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<tr>
<td>Caval</td>
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<td>13</td>
<td>3</td>
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<td>17</td>
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<td>Atrial</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>2</td>
<td></td>
<td>10</td>
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<td>Makeshift shunts</td>
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<td>4</td>
<td>0</td>
<td>0</td>
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<td>4</td>
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<td>Devascularizations</td>
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<tr>
<td>Sugiura-Futagawa</td>
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<tr>
<td>Abdominal</td>
<td>0</td>
<td>40</td>
<td>133</td>
<td>78</td>
<td></td>
<td>251</td>
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<tr>
<td>Thoracic</td>
<td>0</td>
<td>33</td>
<td>93</td>
<td>35</td>
<td></td>
<td>161</td>
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<tr>
<td>1-Stage</td>
<td>0</td>
<td>6</td>
<td>14</td>
<td>3</td>
<td></td>
<td>23</td>
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<tr>
<td>Stapled</td>
<td>0</td>
<td>0</td>
<td>5</td>
<td>0</td>
<td></td>
<td>5</td>
</tr>
<tr>
<td>Non-Sugiura-Futagawa</td>
<td>10</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td></td>
<td>14</td>
</tr>
<tr>
<td>Splenectomy</td>
<td>13</td>
<td>9</td>
<td>0</td>
<td>0</td>
<td></td>
<td>22</td>
</tr>
<tr>
<td>Selective surgery</td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td></td>
<td>6</td>
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</table>
# Table 2. Comparison of the Main Types of Procedures*

<table>
<thead>
<tr>
<th>Variable</th>
<th>DSRS</th>
<th>LDMS</th>
<th>Mesorenal or Mesocaval</th>
<th>Proximal Splenorenal</th>
<th>Portacaval</th>
<th>Sugiura-Futagawa</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>296</td>
<td>37</td>
<td>27</td>
<td>38</td>
<td>103</td>
<td>27 One-stage and 251 abdominal-stage</td>
</tr>
<tr>
<td>Operative mortality, %</td>
<td>5/3†</td>
<td>3</td>
<td>10</td>
<td>14</td>
<td>11</td>
<td>6/2†</td>
</tr>
<tr>
<td>Rebleeding, %</td>
<td>6</td>
<td>15</td>
<td>5</td>
<td>11</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>Encephalopathy, %</td>
<td>8‡</td>
<td>38</td>
<td>40</td>
<td>40</td>
<td>43</td>
<td>4‡</td>
</tr>
</tbody>
</table>

*DSRS indicates distal splenorenal shunt; LDMS, low-diameter mesocaval shunt.
†Past 5 years.
‡Significant difference (P<.05).

- 508 patients with portal hypertension treated surgically between 1991-2002
- 425 males and 83 females
- Mean age 40.8 years
Etiology

- Posthepatitic cirrhosis 92%
- Biliary cirrhosis 4.3%
- Alcoholic cirrhosis 1.6%
- Extrahepatic portal obstruction 1.2%
- Schistosomiasis <1%
- Idiopathic <1%
Surgical Intervention

- Portoazygous disconnection 50%
- Nonselective shunt 33%
- Selective shunt 12%
Nonselective Portosystemic Shunts

<table>
<thead>
<tr>
<th>Types of operation</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Portacaval shunt (PCS)</td>
<td>14</td>
</tr>
<tr>
<td>Portacaval shunt with prosthetic H-graft (PCS-H)</td>
<td>25</td>
</tr>
<tr>
<td>Mesocaval shunt (MCS)</td>
<td>66</td>
</tr>
<tr>
<td>Inferior mesocaval shunt (IMCS)</td>
<td>26</td>
</tr>
<tr>
<td>Splenorenal shunt (SRS)</td>
<td>12</td>
</tr>
<tr>
<td>Juxtal splenocaval shunt (SCS)</td>
<td>16</td>
</tr>
<tr>
<td>Branch of mesenterico-caval shunt (BMCS)</td>
<td>5</td>
</tr>
<tr>
<td>Inferior meso-left renal vein shunt</td>
<td>2</td>
</tr>
<tr>
<td>Colonic media vein-caval shunt</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>167</strong></td>
</tr>
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</table>
# Selective Shunts

## Table 2 Patients treated with selective shunt (SS)

<table>
<thead>
<tr>
<th>Types of operation</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Distal splenocaval shunt (DSCS)</td>
<td>42</td>
</tr>
<tr>
<td>Distal splenorenal shunt (DSRS)</td>
<td>16</td>
</tr>
<tr>
<td>Coronary vein-caval shunt</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td>62</td>
</tr>
</tbody>
</table>
## Change In Free Portal Pressure

<table>
<thead>
<tr>
<th>Types of operation</th>
<th>Cases</th>
<th>Pre-operation (kPa)</th>
<th>Post-operation (kPa)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Portaazygous disconnection (PAD)</td>
<td>81</td>
<td>3.78±0.50</td>
<td>3.26±0.57</td>
</tr>
<tr>
<td>PSS (diameter of anastomotic stoma in 8–12 mm)</td>
<td>52</td>
<td>4.29±0.80</td>
<td>3.11±0.63</td>
</tr>
<tr>
<td>PSS (diameter of anastomotic stoma in 6–8 mm)</td>
<td>42</td>
<td>3.91±0.48</td>
<td>3.12±0.46</td>
</tr>
<tr>
<td>Selective shunt (SS)</td>
<td>62</td>
<td>3.85±0.59</td>
<td>3.26±0.45</td>
</tr>
<tr>
<td></td>
<td>Hepatic Encephalopathy</td>
<td>Operative Mortality</td>
<td></td>
</tr>
<tr>
<td>----------------</td>
<td>-------------------------</td>
<td>---------------------</td>
<td></td>
</tr>
<tr>
<td><strong>PAD</strong></td>
<td>3.9%</td>
<td>5.8%</td>
<td></td>
</tr>
<tr>
<td><strong>PSS</strong></td>
<td>9.6%</td>
<td>4.2%</td>
<td></td>
</tr>
<tr>
<td><strong>SS</strong></td>
<td>4.8%</td>
<td>4.8%</td>
<td></td>
</tr>
</tbody>
</table>
“Different patients at different stages in their disease may require different therapies”

- Patient and surgeon dependent
- TIPS and surgical shunts decrease hemorrhage but do not increase overall survival
- Beta blockade is the only proven treatment investigated with randomized controlled studies