Rex Shunts: 
the role of IR

James Donaldson, MD
Lurie Children’s Hospital of Chicago
IR and Rex Shunts

- Portal Hypertension
- The Rex Shunt
- Pre-operative evaluation
- IR
- Results
Portal Hypertension in Children

- ...with progressive hepatic dysfunction
  - Biliary diseases
  - Parenchymal diseases
  - Hepatic venous outflow obstruction

- ...without progressive hepatic dysfunction
  - Hepatic fibrosis
  - Klatzkin’s disease (angiodysplasia)
  - Splenic vascular anomaly
  - Portal vein obstruction
    - Portal vein anomalies
    - Cavernous transformation
Extra-hepatic portal vein obstruction

- Etiology usually unknown
- History of UVC
- Prematurity
- PV thrombosis
  - Gastroenteritis
  - Dehydration
Management portal hypertension without hepatic dysfunction

- Serial sclerotherapy
- Partial splenic embolization
- Splenectomy
- Surgical management
Surgical Management

- Selective Shunt
  - Warren (Spleno-renal)
  - Rex (Meso-Rex)

- Non-selective
  - Mesocaval

- Segura (gastric venous devascularization)

Rex Shunt

- Decompress portal hypertension
- Restores mesenteric/portal blood flow to liver
- Growth increase (BMI)
- Improve hepatic function
  - Ammonia
  - Neurocognitive improvement

SAM Question #1

The Rex Shunt is appropriate for which of the following:

1. Abernethy malformation Type I
2. A complication of umbilical venous catheterization
3. Veno-occlusive disease
4. Budd Chiari
5. Alagille syndrome
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The following are advantages of the Rex shunt over a Warren shunt except:

1. Reduced bleeding from esophageal varices
2. Reduced incidence of hepatopulmonary syndrome
3. Improved portal venous blood flow
4. Improved cognition
5. Improved BMI
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3. Improved portal venous blood flow
4. Improved cognition
5. Improved BMI
Pre-operative Evaluation

- US
- CTV
- MRV
- Direct PV puncture
- Transjugular wedged portal venography
Transjugular Wedged catheter

TJ wedged hepatic vein portography

Portal Vein
Multiple wedged sites
Rex Shunt Surgery

- Right IJ vein graft
- Dissection of the Rex recessus
- Access of the left PV
- Appropriate mesenteric vein access site
Patent shunt
Pre and Post

+ 12 months
Increased liver volume
1 day post-op - Occluded shunt
17% patients develop stenosis at PV end

Surveillance of Rex Shunts
Surveillance of Rex Shunts
Surveillance of Rex Shunts

- MRV
Surveillance of Rex Shunts

- **Clinical**
  - Splenomegaly
  - Thrombocytopenia
  - GI bleeding

- **Imaging**
  - US
  - CTV
  - MRV
Rex Interventions

- Transhepatic PV access
- Right-sided
- US-guided
- 22 gauge needle
Introduction of vascular sheath

Heparin

Probe PV to find Rex

Identify stenosis

Pressure gradient

> 5 mm Hg
Pre and post angioplasty

6 mm balloon
Evaluation of shunt
Recurrent stenosis post stent
Kissing stents
IR management

* 15 patients with stenosis
  * 9 (60%) primary venoplasty
  * 3 (20%) required multiple procedures
  * 3 (20%) failed interventions – Rex abandoned

* 27 Interventional procedures
  * 17 venoplasties
  * 4 cutting balloons
  * 5 stents
  * 1 venoplasty with thrombolysis

Results

- Pressure gradient reduced 11 to 5 mm Hg
- Platelets increased 97K to 165K
- Ammonia reduced 44 to 24 µmol/L
Slowly withdraw sheath

Parenchymal tract

Back table demo
1 major complication
Rex Shunts
Lurie Children’s Chicago
1997-2012

- 143 patients attempted Rex shunts
- 104 Patent to date
* Indications for IR investigation of a Rex shunt include all except:

1. Thrombocytopenia
2. A suspected stenosis by US, CT, or MR imaging
3. Splenomegaly
4. Recurrent UGI bleeding
5. Lower extremity edema
SAM Question #3

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3. Splenomegaly
4. Recurrent UGI bleeding
5. Lower extremity edema
Summary

- The Rex shunt is an ideal surgical option for extra-hepatic PV obstruction
- Pre and post operative imaging is challenging but vitally important for the success of a Rex shunt program
Summary

- IR plays a significant role in the Rex shunt program
- Percutaneous interventions are challenging but can be done safely and effectively in this patient population
References

