

# Specificities of PVT in children

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# Specificities – in patients with healthy liver

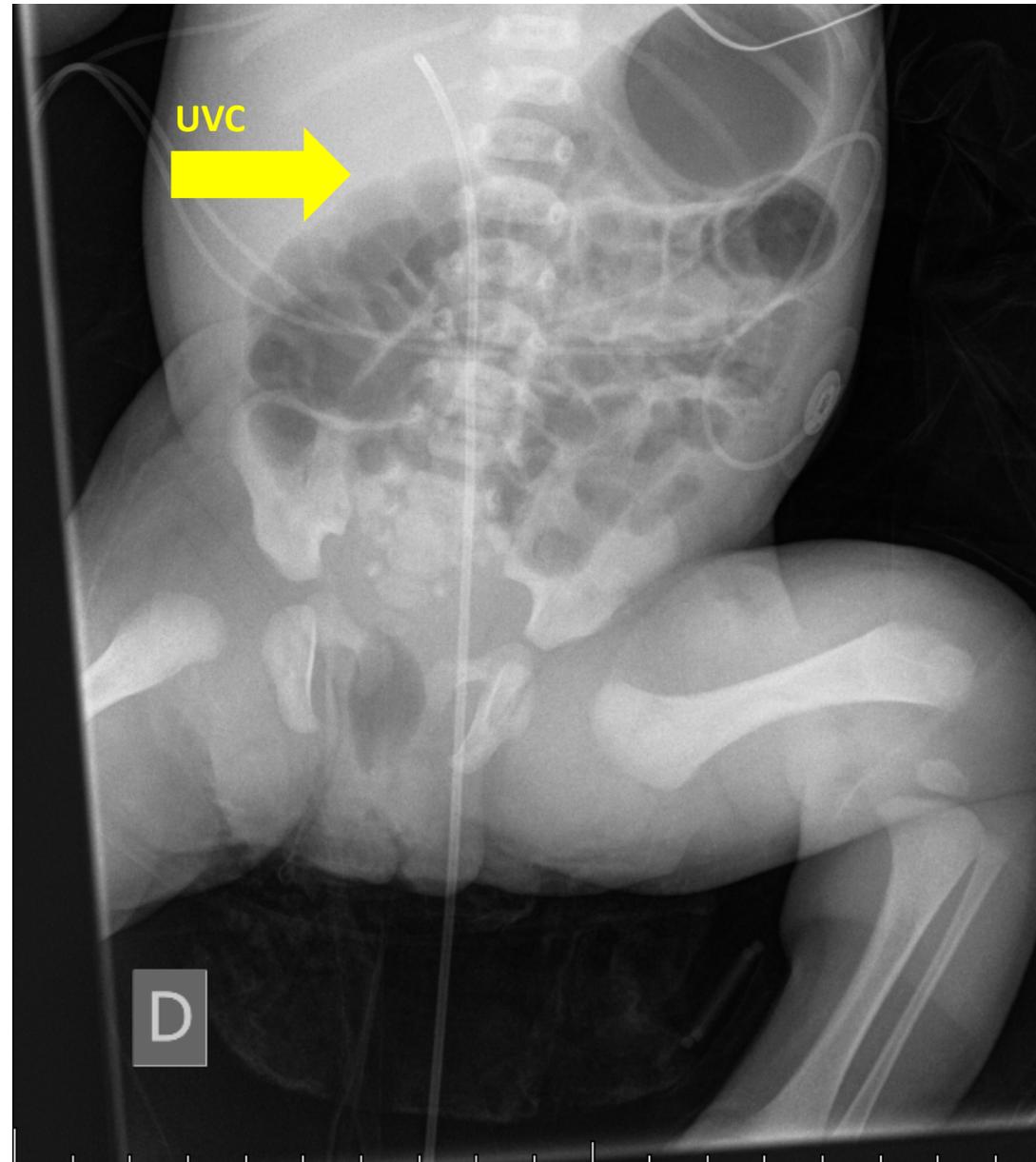
- Etiology
- Presentation
- Management

# Etiology

- Umbilical venous catheter (UVC)
- Others
  - Sepsis (abdominal)
  - Transfusion
  - Sickle cell anemia
  - Splenectomy
- Rarely: thrombophilia

**PVT in cirrhosis very rare**

**Exception: congenital hepatic fibrosis**



# Specificities

- Etiology
- **Presentation**
- Management

# Presentation

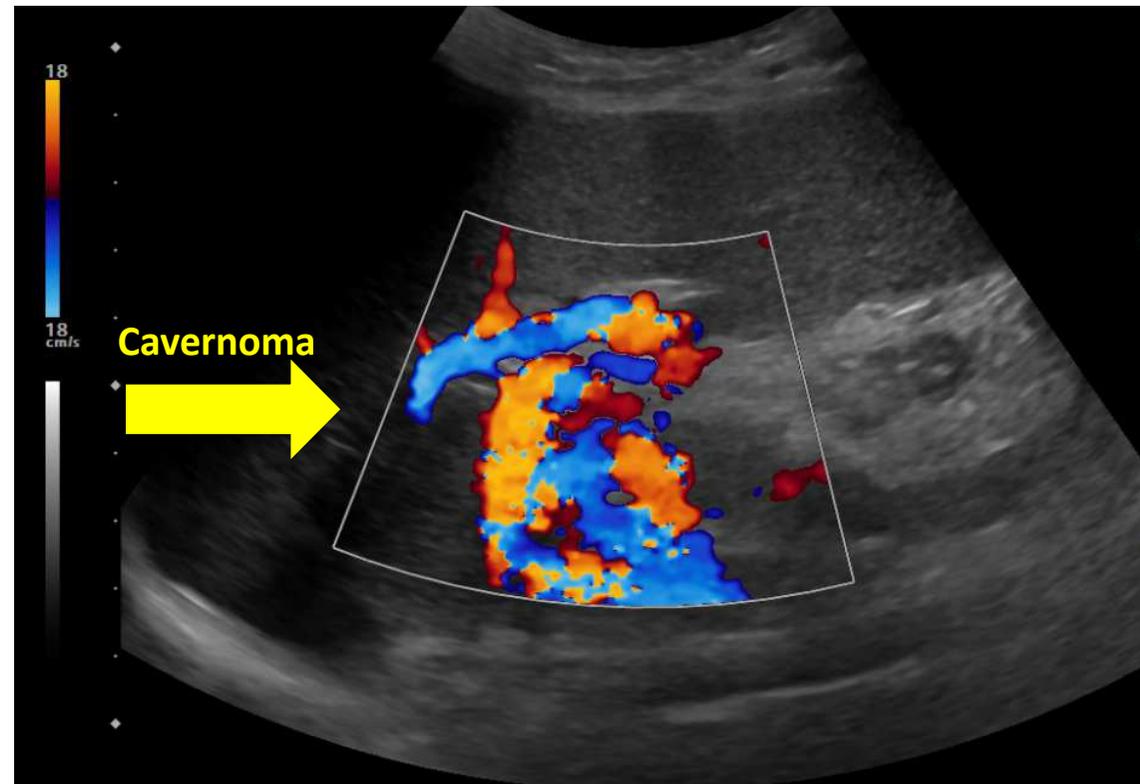
- Isolated splenomegaly

## History

- tracheo-oesophageal fistula
- UVC at birth

Incidence reported 0-43% with UVC

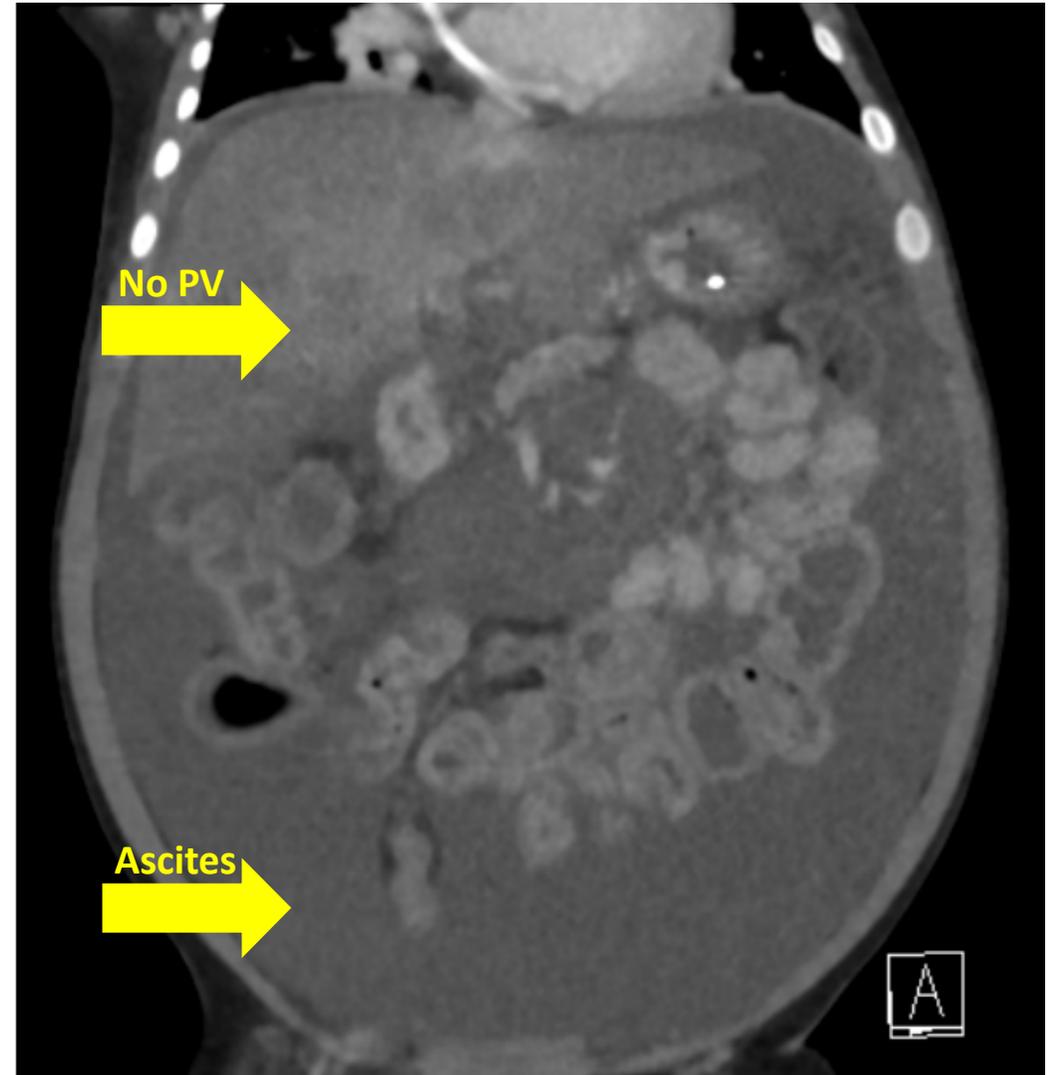
Most common site of thrombosis:  
umbilicoportal confluence (Rex)



# Presentation

- Isolated splenomegaly
- Ascites

**History** abdominal sepsis & persistent fever  
→ Presumed dx of **portal thrombophlebitis**



## Presentation - other

- Rectal varices (hemorrhoids do not exist in children)
- Pancytopenia
  - Suspected marrow failure
- Liver nodules
- **Neonatal Doppler US**



# Management – acute vs chronic

- Anticoagulation (systemic)
- Catheter directed thrombolysis\*

**Acute**

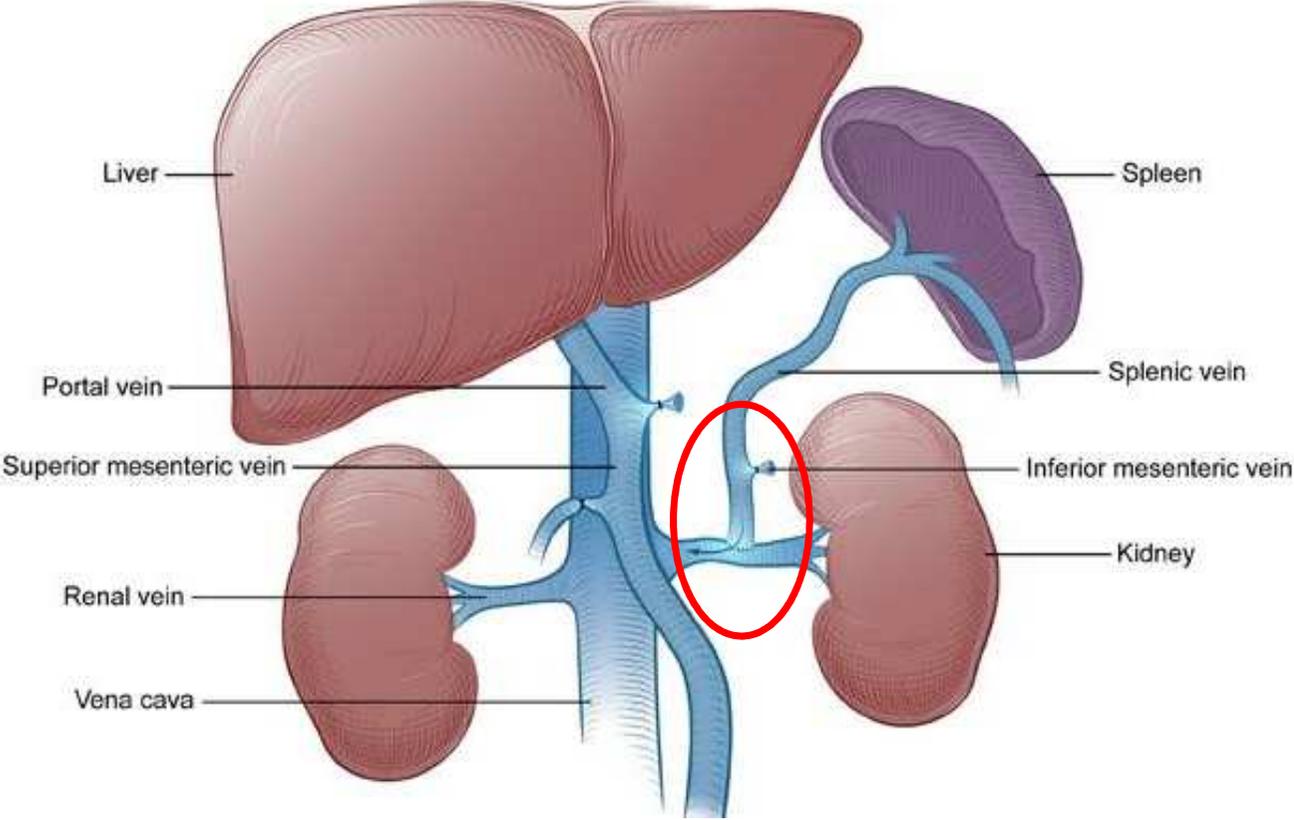
- Watchful management if no complications
- Portal vein recanalization\*\*
- Surgical bypass
  - MesoRex – consider pre-emptively
  - **Splenorenal** – if complications

**Chronic**

\*\*Franchi-Abella, personal communication

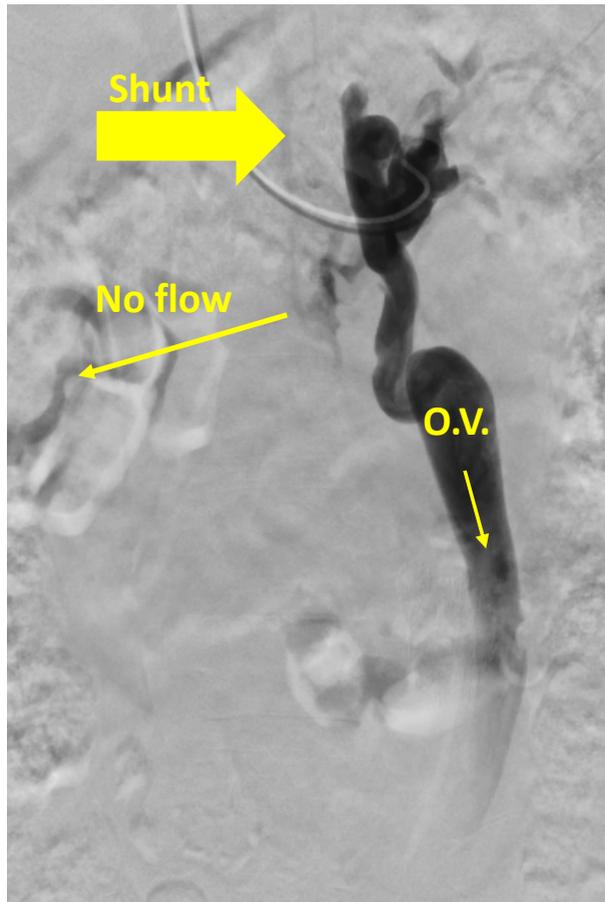
\* Koo et al WJG 2018

# Distal splenorenal shunt



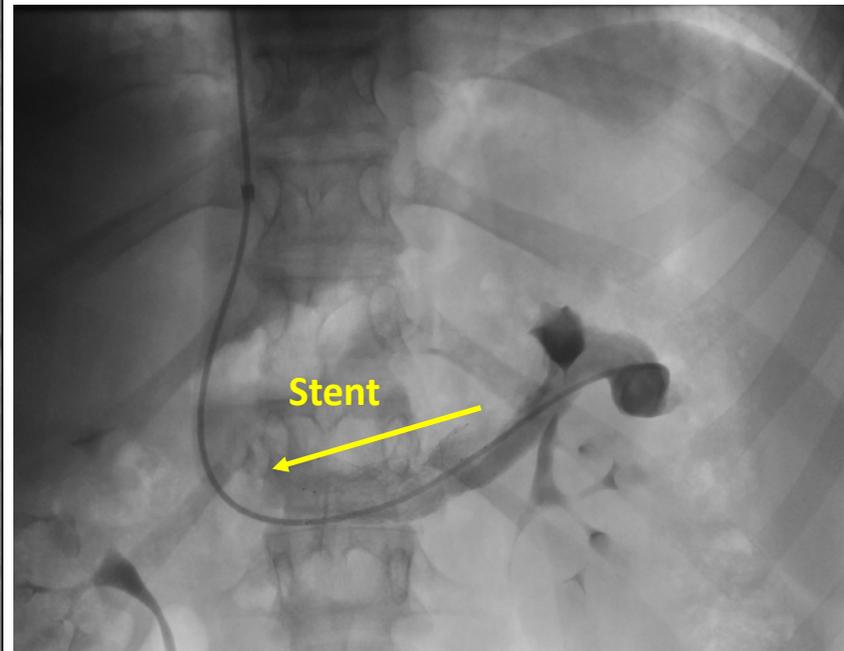
Protective of native liver

# Splenorenal shunt – challenges in children (1)



## Growth:

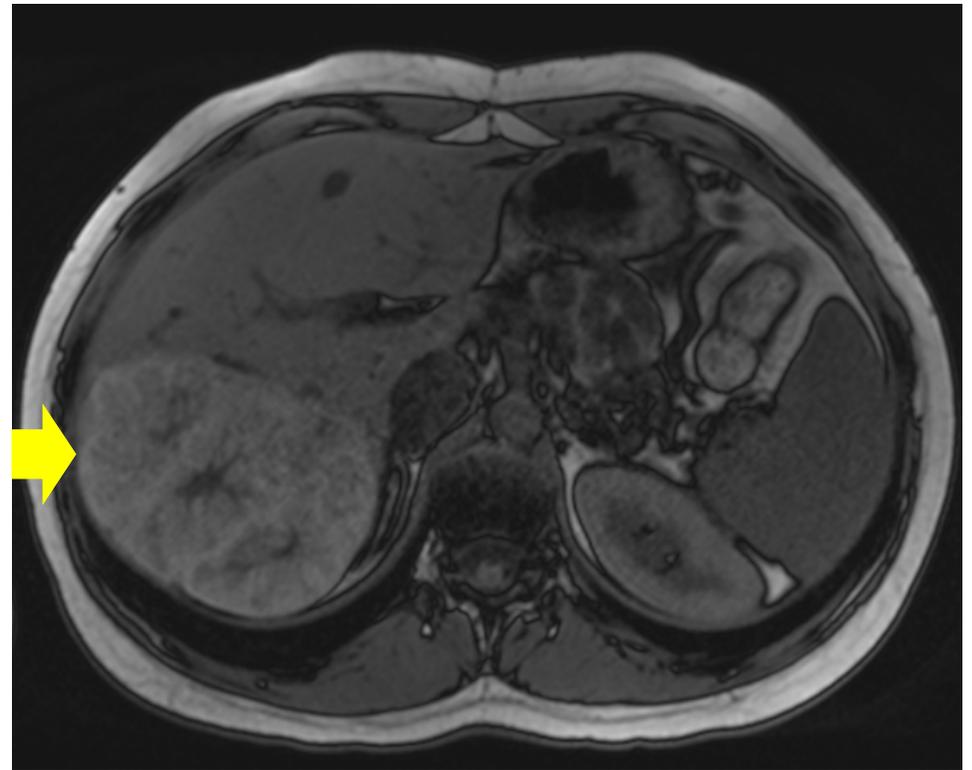
- shunt at age 2
- recurrence of PHT age 14



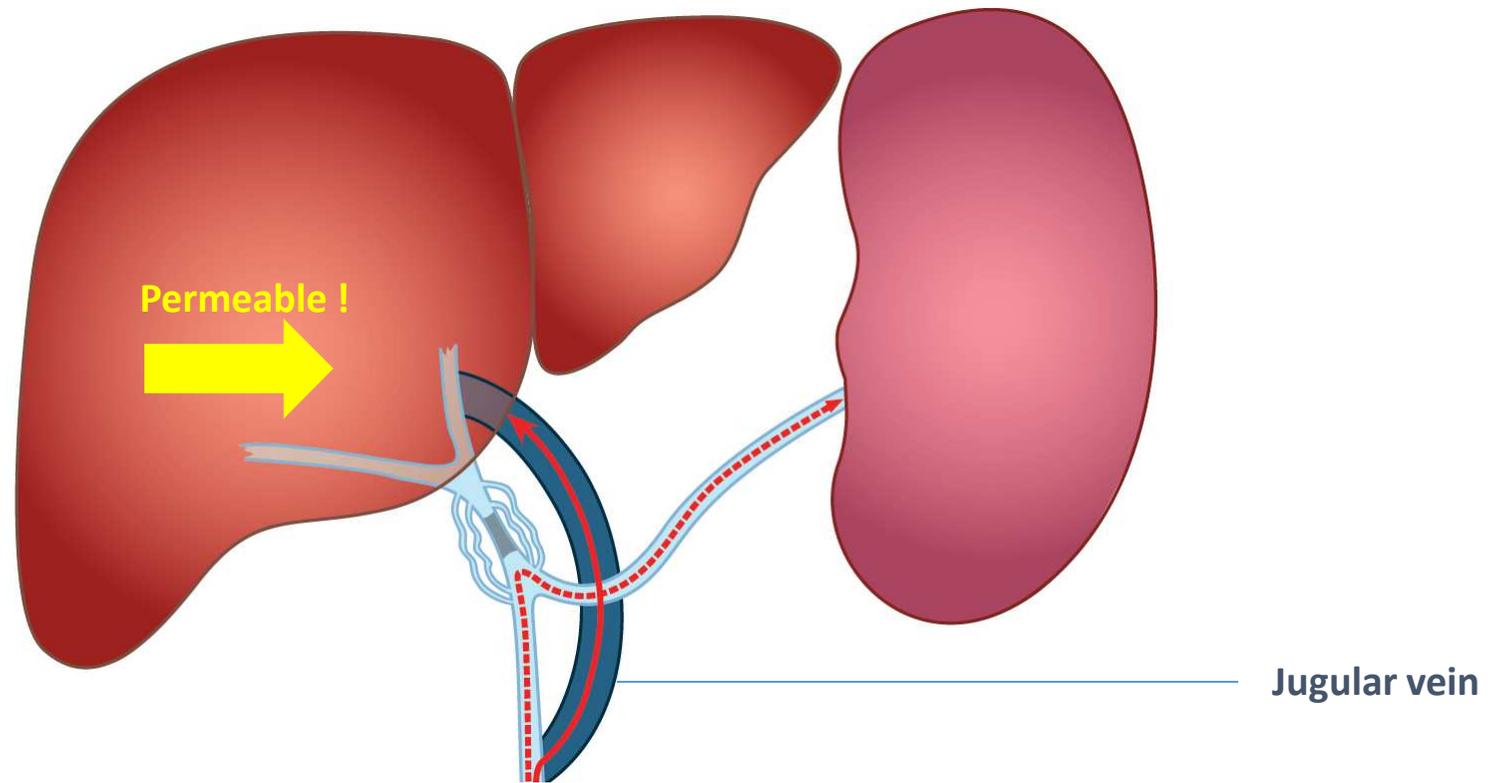
## Challenges (2) in young adults

### Complications of portosystemic bypass

- Hepatopulmonary syndrome ?
- Liver nodules
- Entering university
- Will transition to adult services

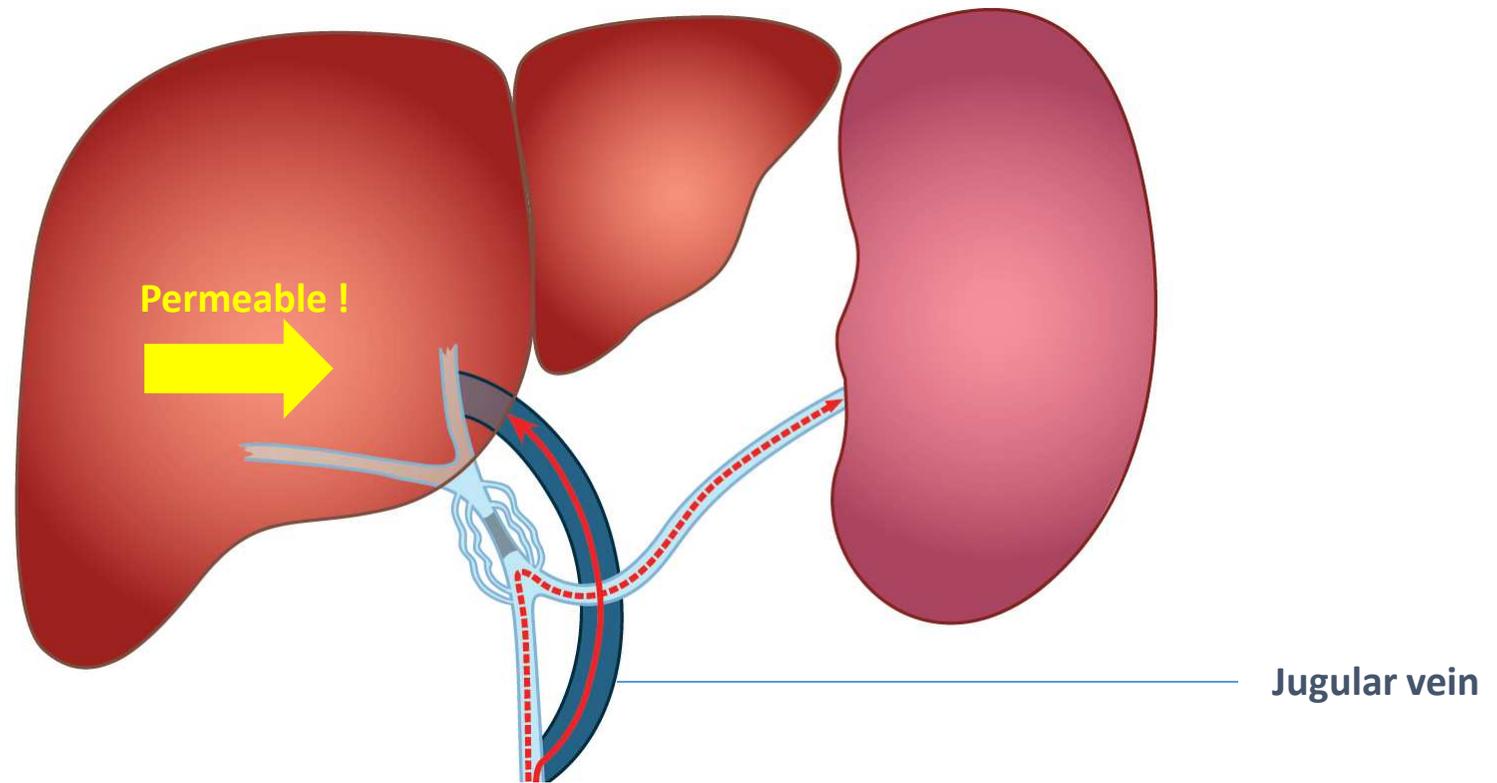


# MesoRex bypass



Retrograde portal venogram essential to assess permeability

# MesoRex bypass



Recommended when feasible – do not wait for complications

# Long term outcomes

## **Surgically Restoring Portal Blood Flow to the Liver in Children With Primary Extrahepatic Portal Vein Thrombosis Improves Fluid Neurocognitive Ability**

Cara L. Mack, MD<sup>a</sup>, Frank A. Zelko, PhD<sup>b</sup>, Joan Lokar, RN<sup>a</sup>, Riccardo Superina, MD<sup>c</sup>, Estella M. Alonso, MD<sup>a</sup>, Andres T. Blei, MD<sup>d</sup>, Peter F. Whittington, MD<sup>a</sup>

**Other signs and symptoms of portal hypertension regress....  
but story still developing**

# In summary – PVT in children

- **Healthy** liver
- **Rex** permeability is key in management
- Surgical portosystemic bypass protective native liver
  - Long term follow up required
    - Growth
    - Complications of portosystemic bypass

# Future directions

- **Prevention** in premies
  
- **Treatment**
  - in premies
  - direct recanalization
  - early meso-Rex
  
- Promoting survival with **native liver**



Portal hypertension



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<https://csfe.hug.ch/en/portal-hypertension-symptom>



# Risk factors for PVT in UVC

- **Catheter related**

- Position: portal vein
- Lower placement protective

- **Patient factors**

- Low birth weight
- Low flow state
- Sepsis
- Maternal gestationnal diabetes
- Hypoxia
- Congenital malformations