CPSS Shunts in Neonates: Importance of Early Diagnostic Work-up

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European Society of Pediatric Radiology

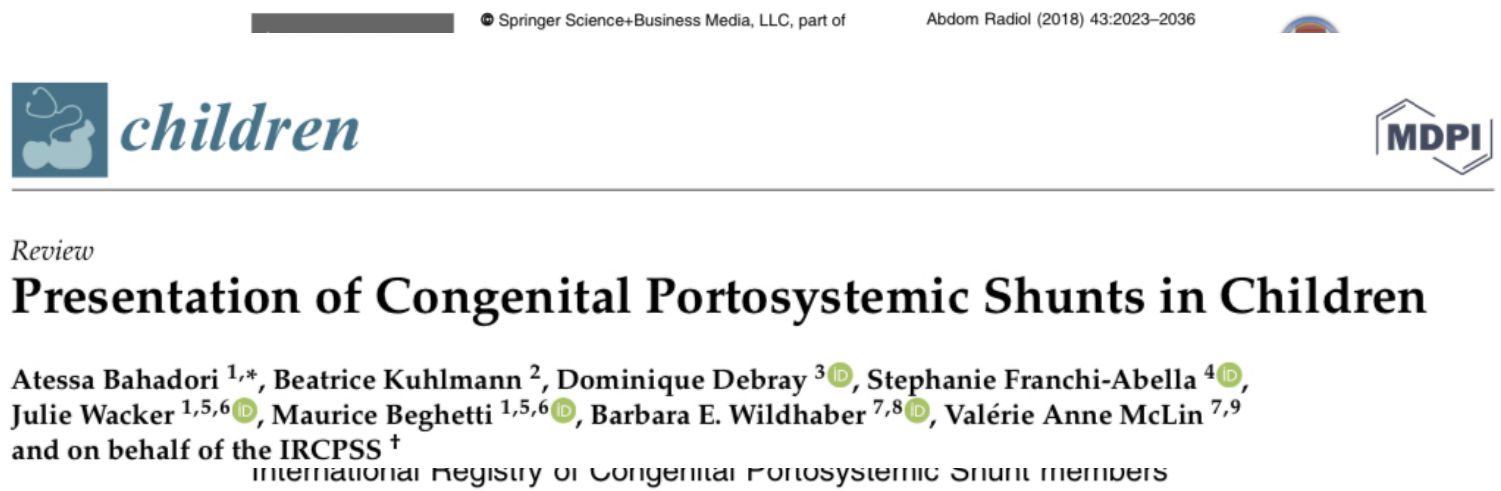
Geneva, Switzerland

ESPR
European Society of Paediatric Radiology

Pubmed

In the last 5 years:

- 240 articles published
- 236 available full text
- 2022: 35 indexed article
- 1 systematic review
- 30 reviews
- Numerous case studies



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children

MDPI

Review

Presentation of Congenital Portosystemic Shunts in Children

Atessa Bahadori [1,*], Beatrice Kuhlmann [2], Dominique Debray [3], Stephanie Franchi-Abella [4], Julie Wacker [1,5,6], Maurice Beghetti [1,5,6], Barbara E. Wildhaber [7,8], Valérie Anne McLin [7,9] and on behalf of the IRCPSS [†]

International Registry of Congenital Portosystemic Shunt members

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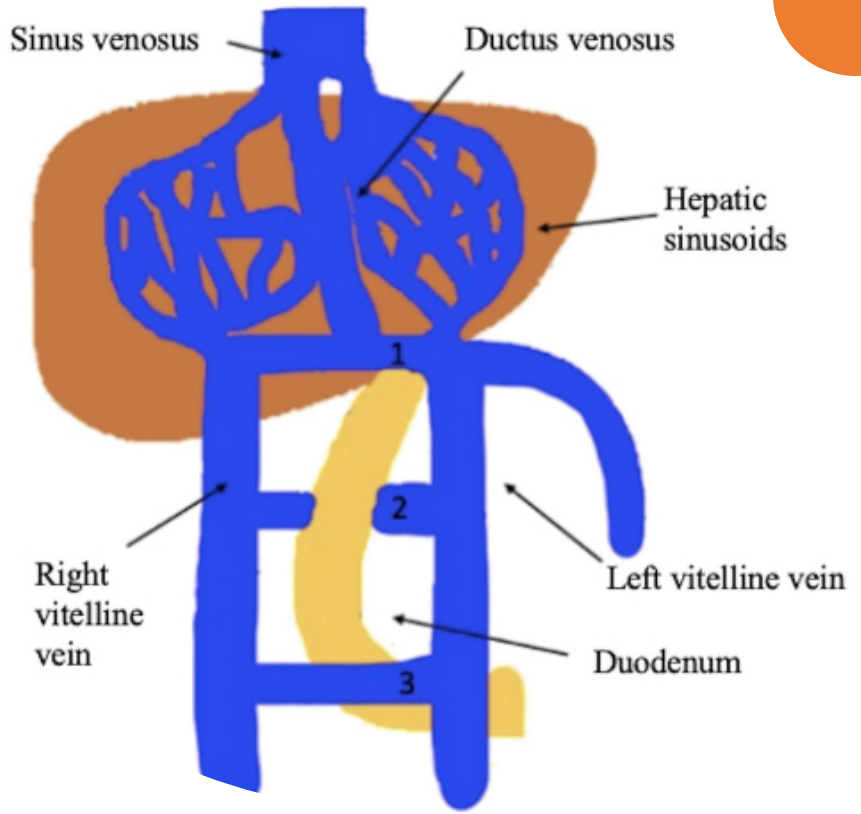
Questions

- Which patients?
- What does the general pediatrian need to know?
- How to image?
- What classification should I use?
- What congenital anomalies and complications exist?
- What do the surgeons and the interventional radiologist need to know?
- What do treatment options look like on imaging?
- What to look for in post treatment control studies?

Definition

- Abnormal vascular connections between the splanchnic and systemic venous circulation, within or exterior to the liver
- 1/6000 fetuses
- 675 live births in Europe/year
- Incomplete remodeling between embryonic and fetal vascular systems
- Low pressure systems
- Major pathophysiologic consequences



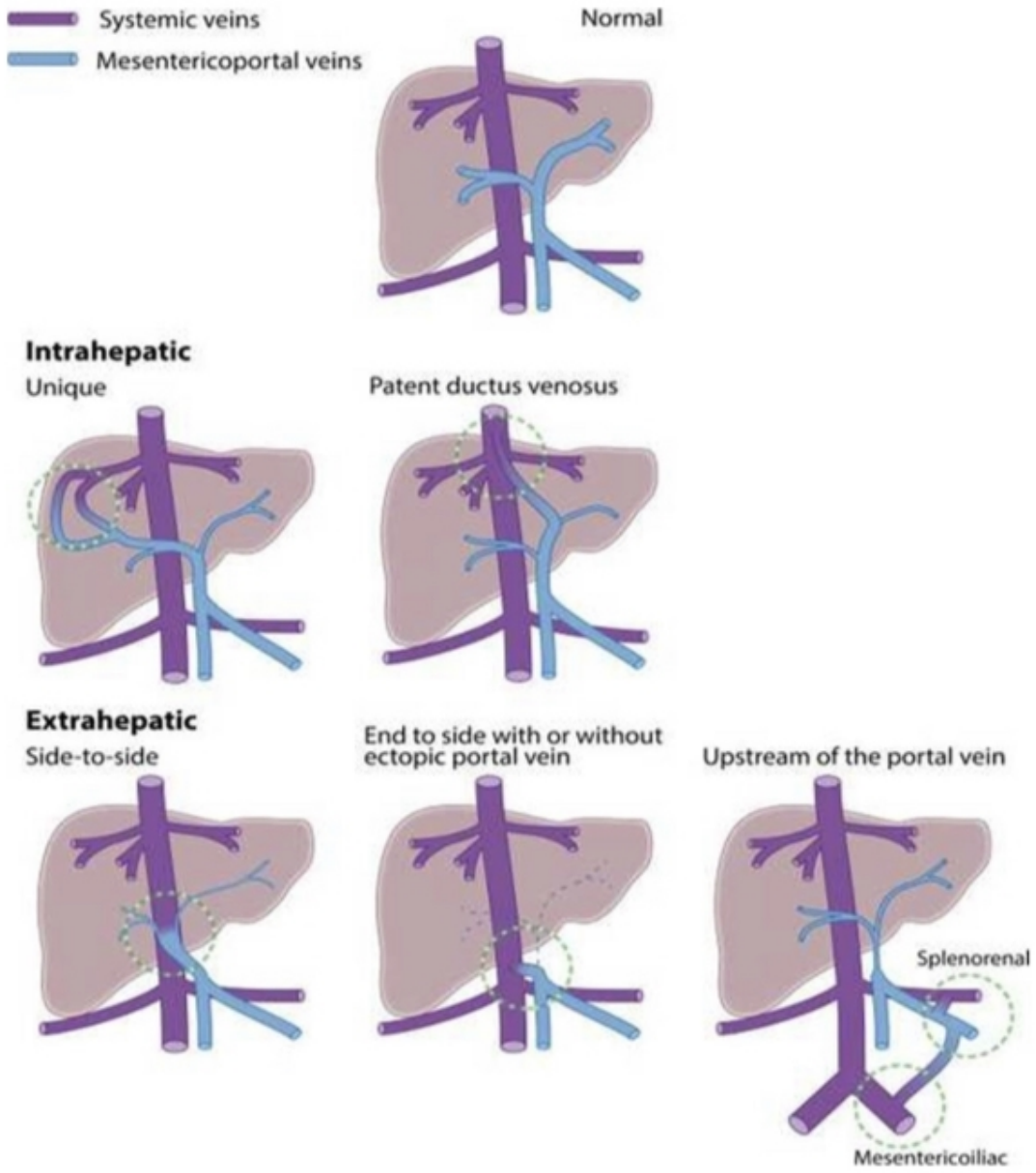
McLin et al. Presentation of Congenital Portosystemic Shunts in Children. Children: 2022:9,243

Kumar P, Bhatia M, Garg A, Jain S, Kumar K. Abernethy malformation: A comprehensive review. Diagn Interv Radiol. 2022;28(1):21-28

Classification: Bictêre

- Classifies shunts based on prognosis
- Type I: extrahepatic portosystemic shunt
- Type II: porto-caval shunt (SSPC, ESPC)
- Type III: Intrahepatic porto-hepatic shunt
- Type IV: Persistent ductus venous
  - (after 1st month of life)

- Shunts that will likely spontaneously close (80% intra-hepatic shunts)
- Shunts that will likely remain open (some intrahepatic shunts, extra-hepatic shunts, patent ductus venous)



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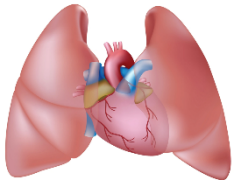
Uchida et al. Long term outcome of liver transplantation for congenital extrahepatic Portosystemicc shunt. Liver Transplantation. 2021: Feb 27 (2): 236-247.

et la. Congenital Portosystemic shunts: diagnosis and treatment. Abdominal Radiology:2018: 43:2023-2036Franchi-Abelli

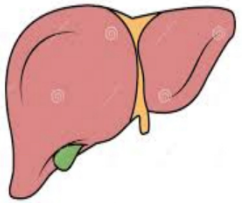
Clinical presentation



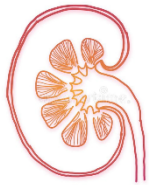
Cardiopulmonary
- **Hepatopulmonary Sx**
- **PA hypertension**
- **Cardiac failure**

Incidental finding on prenatal US
Low birth weight

Neurocognitive
- **Mild impairment**
- ADHD
- **Behavioural** problems
- **Encephalopathy**
- Seizures
- Parkinson-like



Liver
- **Liver tumors**
- **Neonatal cholestasis**
- **Elevated liver function/enzymes**



Renal
- Membrane proliferative glomerulopathy

Hematologic
Coagulopathies

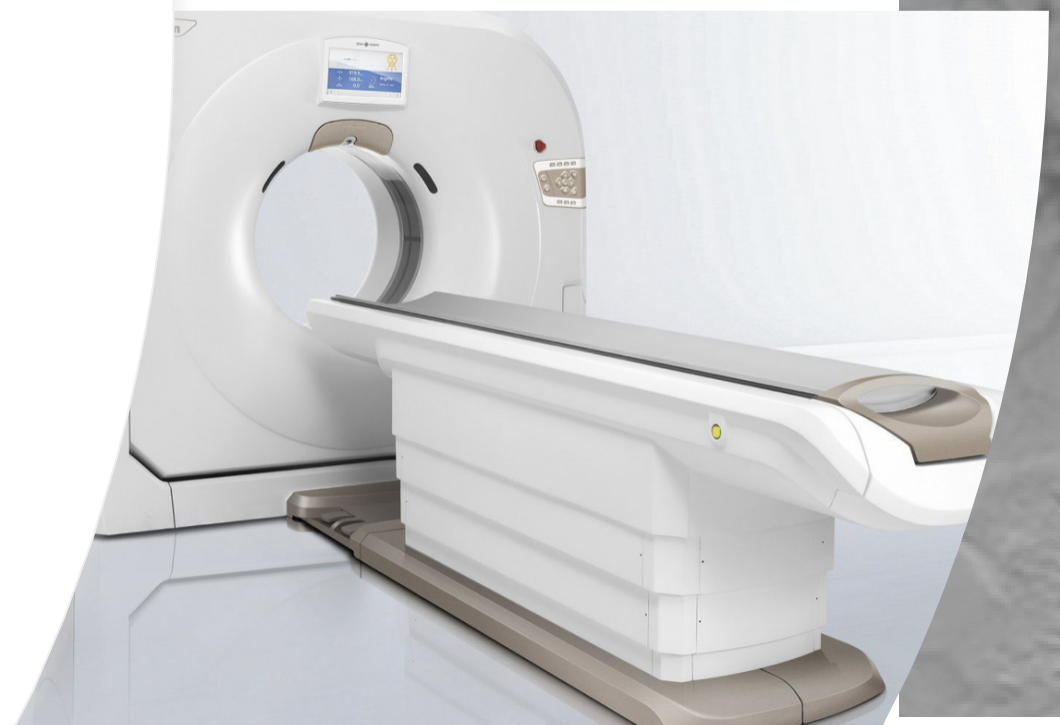
Syndromes
- Caroli
- Goldhenhar
- Down’s
- Turner’s

Endocrine
- Glucosemia
- Galactosemia
- Hyperammonia

Bahadore et al. Presentation of Congenital Portosystemic Shunts in Children. Children. 2022 Feb 11;9(2):243

Imaging of choice

- **Ultrasound**
- CT
- MRI
- Diagnostic angiography



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Initial diagnosis in neonates

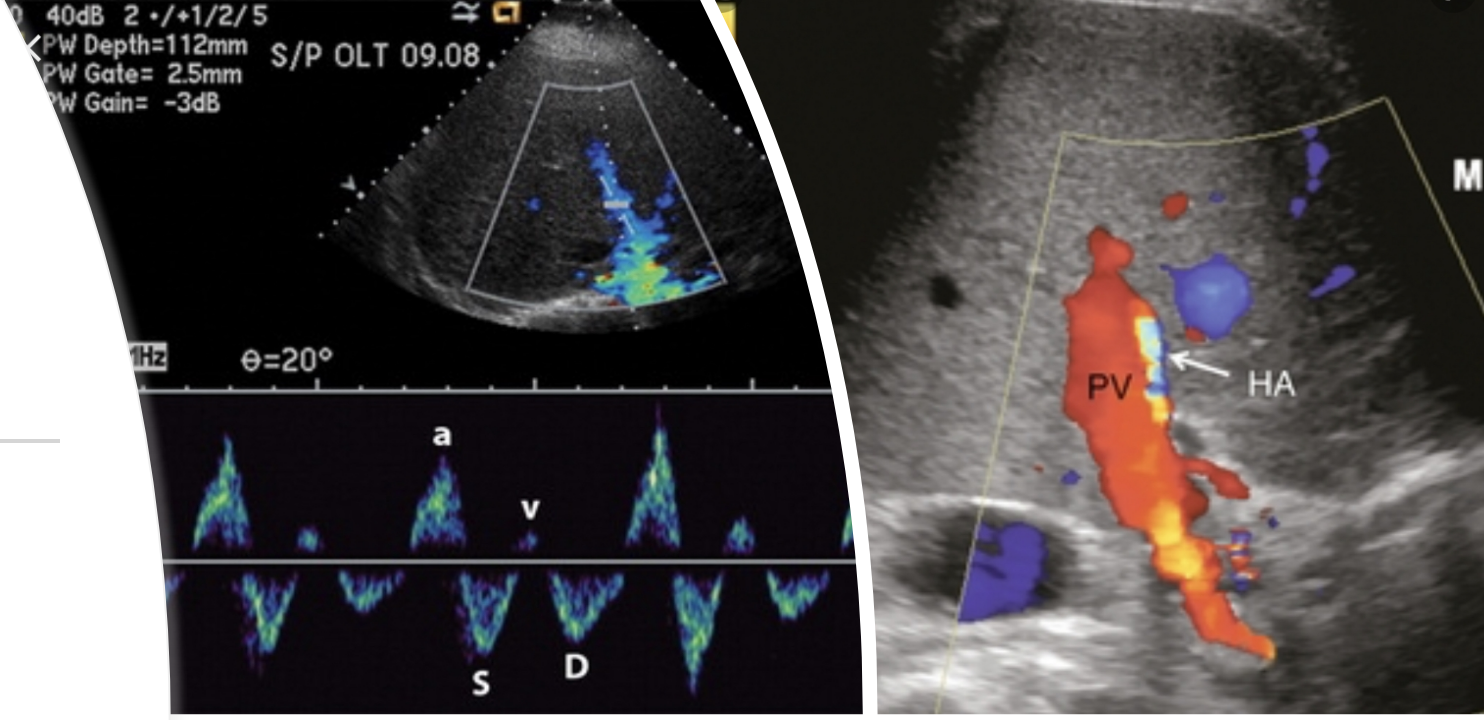
- Basic work-up
  - Clinical exam (malformations, cutaneous hemangiomas)
  - Bloodwork
  - Ultrasound

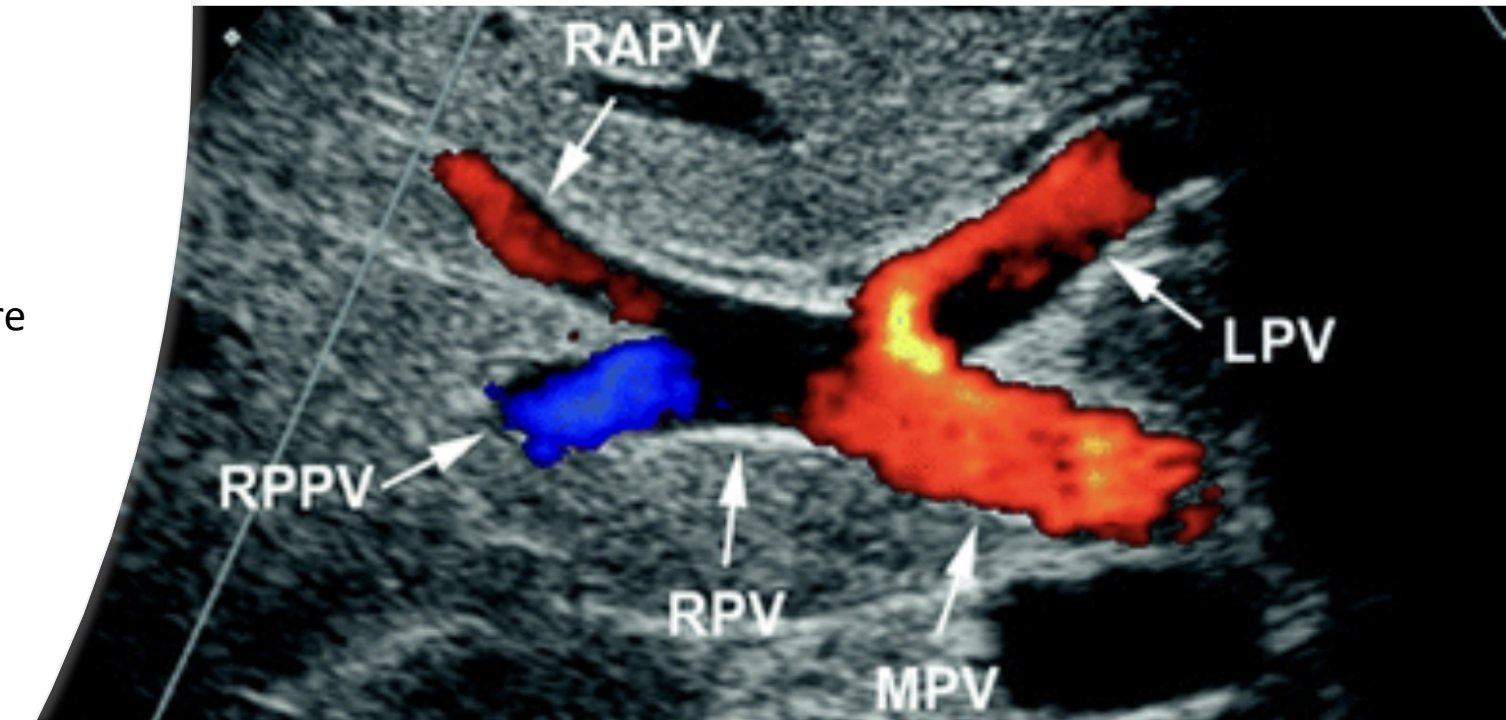
Francois et al. Prenatally diagnosed congenital portosystemic shunts. Journal of Maternal-Fetal &Neonatal Medicine. 2018. Vol 31, No 10, 1364-1368

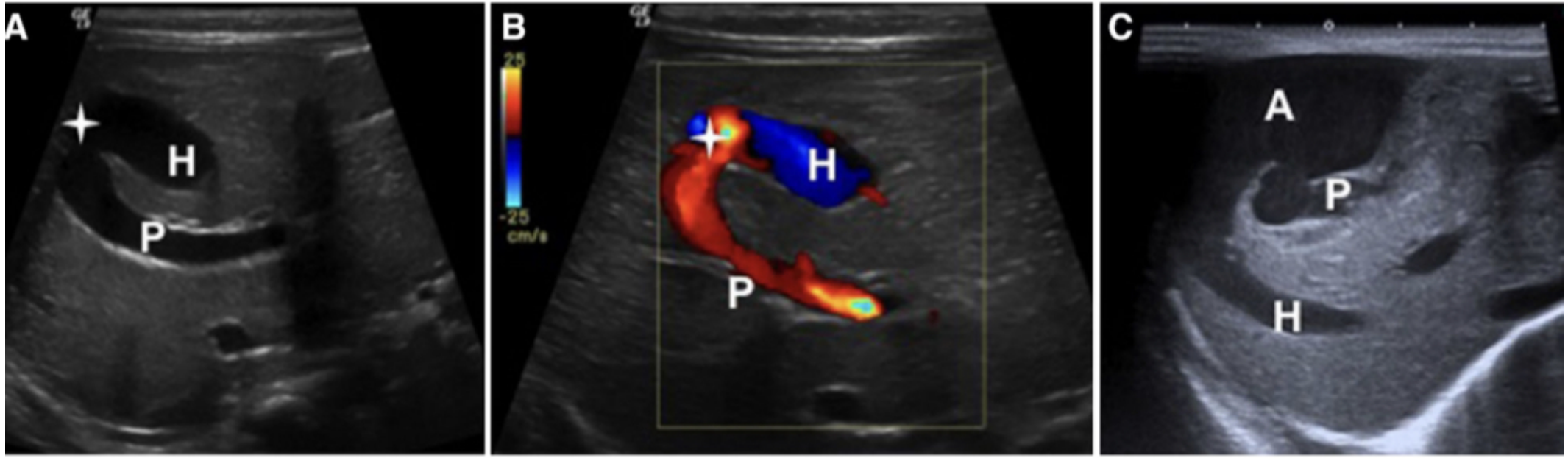
Bahadore et al. Presentation of Congenital Portosystemic Shunts in Children. Children. 2022 Feb 11;9(2):243

Early Ultrasound

- Systematic analysis of the following:
  - Dilated hepatic veins
  - Follow hepatic veins centrally to periphery with Doppler
  - Abnormal vascular angulation
  - Aneuvrysmal vascular formation
  - Persistent ductus venosus
  - Intra-hepatic portal veins: presence & calibre
  - portal vein: dilatation, anomalies, communication (ESPC, SSPC, H-type PC)
  - Elastography



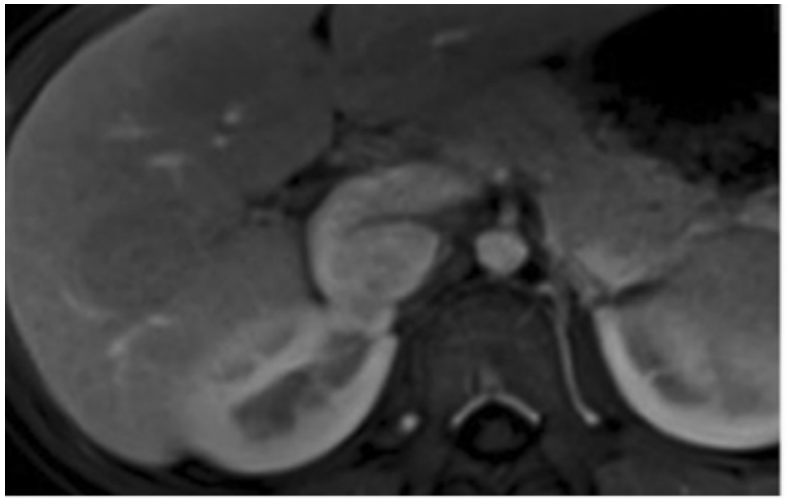




Portohepatic shunt between right portal vein and median hepatic vein. C) Aneurysmal communication between right portal vein and right hepatic vein. Franchi-Abella et al. Congenital Portosystemic shunts: dianosis and treatment. Abdominal Radiology (2018) 43:2023-2036.

Follow-up

- Doppler US and elastography at 1, 3, 6 & 12 months
- If persistent shunt:
  - Liver biopsy: nodules, underlying liver disease
  - CT/MRI: shunt anatomy, work-up for liver masses
  - Cardiac echo: R-L shunt
  - Pulmonary scintigraphy
  - Brain MRI: T1 hyperintense Globus palladi (manganese, glutamine/glutamate deposition)



Franchi-Abella et al. Congenital Portosystemic shunts: dianosis and treatment. Abdominal Radiology (2018) 43:2023-2033

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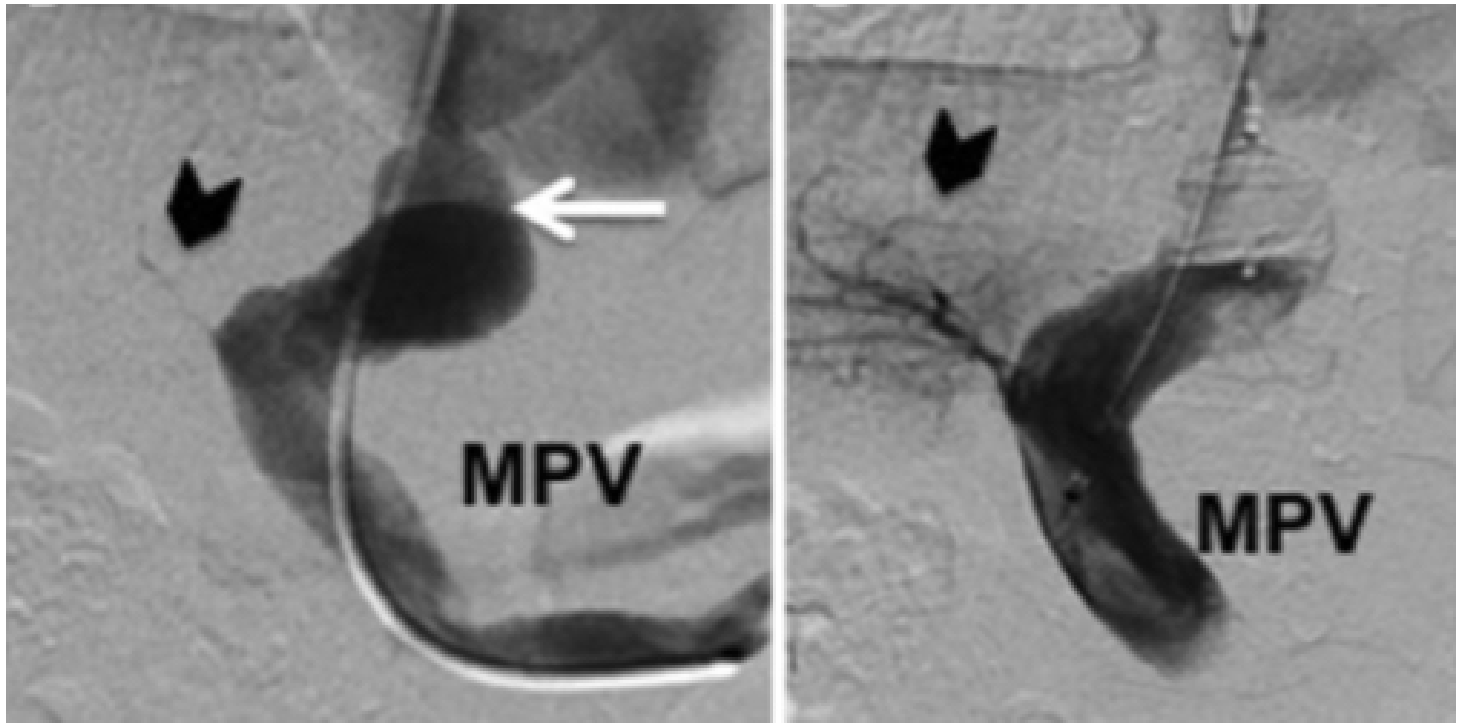
Treatment options

Preventative closure of persisting shunt after the age of 2yrs is recommended

- Angiographic closure
- Surgical ligation
- Liver transplant (very few cases)



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Post-treatment imaging

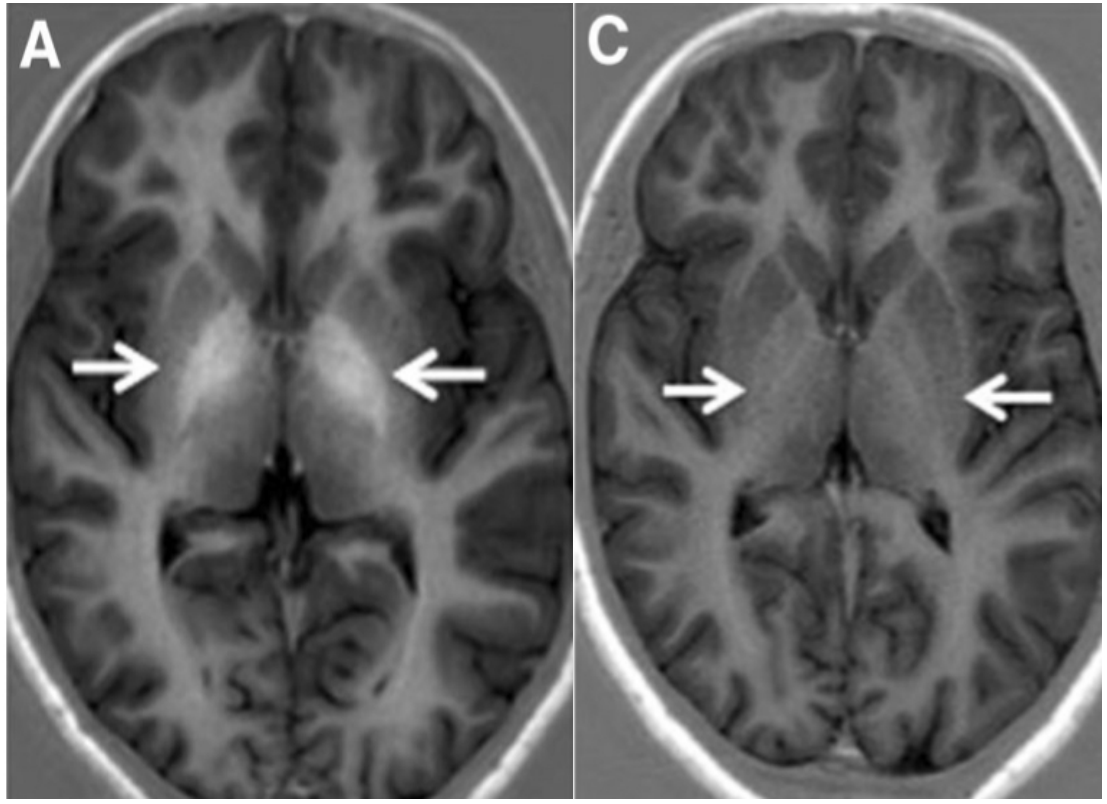
Close follow-up by ultrasound:

- Look for post treatment thrombosis
- Identify intra-hepatic portal venous system
- Identify embolization material and analyze permeability on 2D and doppler study
- Spectral doppler
- Elastography

- If clinical or radiologic suspicion: further work-up (CT and/or angiography)

Outcomes

- Restored intra-hepatic portal flow
- Complete or partial regression of benign liver masses
- Regression and stabilization of complications:
  - Neurological
  - Pulmonary
  - Cardiac
  - Renal



Bernard et al. Complications of congenital Portosystemic shunts in children: therapeutic options and outcomes. J Pediatr Gastroenterol Nutr. 2010. Sept: 51 (3): 322-30
et la. Congenital Portosystemic shunts: diagnosis and treatment. Abdominal Radiology:2018: 43:2023-2036Franchi-Abelli

Key messages

- Children with multiple and unrelated symptoms?
  - Consider CPSS
- Initial radiologic work-up:
  - Doppler ultrasound of the liver
- Secondary radiologic work-up for persistent shunts:
  - Abdominal CT/MRI
  - Angiography
  - Brain MRI

Symposium:
Taking Congenital Porto-systemic Shunt to the next level-2nd expert meeting

- International Registry of Congenital Porto-Systemic shunts (IRPSS)
- July 1-2, 2022
- Geneva, Switzerland
- On-site & online

- www.hug.ch/en/symposium-taking-cpss-to-next-level
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