Congenital Portosystemic Shunts: Variable Clinical Presentations
Requiring A Tailored Endovascular or Surgical Approach

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Background

- Congenital portosystemic shunts (CPSS) are rare developmental anomalies resulting in diversion of portal flow to the systemic circulation with an incidence of approximately 1:30,000 births. If the shunts are persistent or large, they may lead to long-term complications.
- CPSS are divided into intrahepatic and extrahepatic categories.
- Many shunts spontaneously close by 1 year of age. Persistent shunts require intervention; Shunt morphology and flow dynamics help determine approach for closure.
- Our goal is to show the various clinical presentations, treatments, and outcomes in a subset of pediatric patients at Children’s Hospital of Colorado (CHCO).

Methods

- From 2016 - 2021, 5 pediatric patients (Age: 33 weeks to 6 years) were diagnosed with CPSS at CHCO.
- Retrospective data collection was performed independently by two radiology residents (EBR and GAJ) under the supervision of an attending radiology physician (AA).
- Multidisciplinary data coororation was subsequently performed.

Results

Case 1
34 week old male with intrahepatic shunt resulting in hepatic hydrops with hyperperfusion, cholestasis, hyperinsulinemia and cardiomegaly. Resolution of hypoglycemia after shunt closure. Long, narrow morphology; Endovascular closure performed.

Case 2
33 week old female with intrahepatic hemangiomatosis, causing hepatic dysfunction and severe pulmonary hypertension causing decreased hepatic clearance of vasoactive substances. Decreased pulmonary hypertension with shunt closure. Multiple shunts; Endovascular closure performed.

Case 3
2 year old female with biopsy-proven progressive benign hepatic lesions and symptomatic hypoglycemia, diagnosed with extrahepatic shunt. Resolution of hepatic lesions seen after shunt closure. Short, wide morphology; Staged surgical shunt ligation performed.

Case 4
6 year old female with intrahepatic shunt manifesting as precocious puberty given hyperandrogenemia due to impaired hepatic sulfation of DHEA. Normalization of testosterone levels seen after shunt closure. Long, narrow morphology; Endovascular closure performed.

Case 5
6 year old female with intrahepatic shunt manifesting as precocious puberty given hyperandrogenemia due to impaired hepatic sulfation of DHEA. Normalization of testosterone levels seen after shunt closure. Long, narrow morphology; Endovascular closure performed.

Conclusions

- Congenital portosystemic shunts are rare developmental anomalies that can have an array of clinical presentations.
- Regardless of clinical presentation if a CPSS persists past 1-2 years of age, closure is recommended.
- This case series helps to highlight the various potential clinical presentations, diagnostic work-up, and approaches to shunt closure.

Implications

- Management of these patients should involve a multidisciplinary team including Interventional Radiology, Surgery, Hepatology and other medical services as the patient’s clinical presentation warrants.
- Earlier closure can help prevent metabolic and neoplastic sequelae of persistent portosystemic shunting.

Disclosures

- None