Anesthetic Implications of Hepatic Disease in Children

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Disclosure

- I receive no financial support from, nor have any commercial relationship with, any drug or equipment product, manufacturers, or vendors that may be mentioned or displayed in the course of this presentation.
Objectives

1. List at least five pediatric diseases with liver pathology
2. Describe the complications of advanced hepatic disease
3. Discuss keys to evaluating patients with hepatic disease
Neonatal Cholestasis

- Impaired Development
- Obstruction
- Infections
- Metabolic Disorders
- Alpha 1 Anti-trypsin
- Gestational Alloimmune Liver Disease
- Cystic Fibrosis
- TPN
Alagille Syndrome

- Reduced bile flow because too few bile ducts within the liver

- Associated diseases
  - CV
    - Pulmonary stenosis, TOF
    - Coarctation of the aorta
  - Spine

- Unique facial features
Autosomal dominant, with variable penetration

1 in 100,000 live births

May not be present at birth

Jaundice
  - Conjugated hyperbilirubinemia (4-10 mg/dL)

Hepatomegaly

Pruritus severe by 6 months

15% need liver transplantation
  - ~2% of all liver transplants
Biliary Atresia

- Most common cause of chronic liver disease in newborns
- 1:10,000 to 30,000 live births
- Most common reason for liver transplantation in children
- Blocked common bile duct between liver and small intestine
Infections

- T: Toxoplasmosis
- O: Other (syphilis, parvovirus)
- R: Rubella
- C: CMV
- H: Herpes
Metabolic Disorders

- Galactosemia
- Tyrosinemia
- Lipid metabolism disorders
  - Niemann-Pick
  - Gaucher Disease
Alpha-1 Antitrypsin

- Protein that protects tissues in the body from being attacked by its own enzymes
- Liver produces abnormal protein and it accumulates within the liver cells
- 1:1,500 to 4500
- Up to 20% will have liver involvement
GALD

- Gestational Alloimmune liver disease
  - Neonatal hemochromatosis
- Maternal reactive IgG that attacks the fetal liver
- IVIG and exchange transfusions
Medications

- Antibiotics
  - Clavulanic acid/amoxicillin
  - Timethoprim-sulfamethoxazole
  - -cyclines (minocycline, doxycycline, tetracycline)
  - Macrolides (erythromycin, azithromycin)
  - Rifampin
Medications (cont.)

- Anticonvulsants
  - Phenytoin
  - Valproic acid

- Anti-inflammatory
  - NSAIDs
  - Acetaminophen

- Others
  - Amiodarone
  - Trazodone
Concentrated Drops

160 mg/5ml vs 80 mg/0.8 ml
Other Toxins

- Ecstasy
- Cocaine
- Solvent sniffing (glue)
- Ma Huang
- Black cohosh root
- Kava
- Amatoxin (from Amanita mushroom)
NAFLD

- More common than originally thought
- Associated with
  - Obesity
  - Medications
  - Genetic factors
- As liver tries to process excess fat, inflammation may lead to NASH
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CNS

- Cerebral Edema
- Increased ICP
- Hepatic Encephalopathy

Management
- Oxygen therapy if needed
- Fluid restrict to 85-90% of maintenance
- Head elevation
- Adequate sedation
- Consider empiric broad spectrum antibiotics
Cardiovascular

- Hyperdynamic circulation
  - LV EF usually normal in children
  - Low SVR
  - Compensatory increase CO

- Shunting
  - Cutaneous, splanchnic, intrapulmonary, portopulmonary, lumbral, and pleural

- Activation of sympathetic nervous system
  - Decreased responsiveness to catecholamine infusions
Pulmonary

- Impaired hypoxic pulmonary vasoconstriction
- Restrictive ventilatory pattern
- Atelectasis
- Pneumonia
- Hepatopulmonary syndrome
- Portopulmonary hypertension
Pulmonary (cont.)

- Hepatopulmonary Syndrome
  - Intrapulmonary vasodilation and shunting
  - Impaired oxygen diffusion
  - Increased A:a gradient

- Portopulmonary Hypertension
  - Portal and pulmonary hypertension
  - Increased pulmonary vascular resistance
Renal

- Renin-angiotensin-aldosterone
  - Impaired
    - Sodium handling
    - Water excretion
    - Urine concentrating ability
  - Prerenal insufficiency (ascites or diuretics)
  - Acute renal failure

- Hepatorenal syndrome
  - Rapid deterioration of renal function associated with profound sodium retention
  - Usually preceded by major event such as GI hemorrhage, sepsis, or surgery
Hematologic

- Anemia: bone marrow suppression and bleeding
- Thrombocytopenia: diminished thrombopoietin production and platelet sequestration in spleen
- Coagulopathy: hepatic synthesis of clotting factors impaired
  - dysfibrinogemia
Coagulopathy

- **Pro-coagulant protein reduction**
  - PT/INR
  - Reflect hepatic production of factors V and VII, which have shortest half lives

- **Anti-coagulant protein reduction**
  - Antithrombin, protein C and protein S

- Net result is surprisingly low bleeding complication rate
Metabolic Issues

- Hypoglycemia
  - Impaired hepatic gluconeogenesis and depleted glycogen stores

- Hypokalemia
  - Dilution from volume overload, ascites, or renal wasting

- Hypophosphatemia
  - Unknown mechanism
1. Extrahepatic Biliary Atresia

2. Other

3. Metabolic Disease (other), hepatoblastoma

4. Acute Hepatic Necrosis, other
   • Acute viral infections
   • Autoimmune hepatitis

5. Alagille Syndrome, TPN

UNOS, September 2012
## Latest Statistics from UNOS

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Cardiovascular

- Possible myocardial involvement with metabolic diseases
  - Auscultation, cyanosis, clubbing
- Possible need for further cardiac evaluation
Pulmonary

- CXR
- RA SpO2
- ABG on RA and 100%

- Associated disease considerations
  - CF
  - Alpha-1 antitrypsin deficiency
Renal

- Cr, BUN, GFR
- Fractional Excretion of Sodium
  - >1% indicates ATN
  - <1% indicates HRS and prerenal azotemia
- Avoid nephrotoxic drugs
  - IV contrast dye, aminoglycosides, nonsteroidal anti-inflammatory agents
Coagulation

- Can be most concerning to anesthesiologist, depending on the procedure
- Assess patient’s clinical status and necessity of procedure
- Discuss risk with patient and/or family
- Be prepared
Summary of Objectives

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References


- www.uptodate.com
Thank You

- Questions?