CONTENTS

1. Focal liver masses
2. Biliary tract pathology
FOCAL HEPATIC MASSES

- Broad spectrum in etiology and imaging appearance
CLASSIFICATION OF HEPATIC TUMORS

- Metastatic
- Hepatoblastoma
- Hepatocellular: HCC, FNH, adenoma
- Mesenchymal: “hemangiomas”, mesenchymal hamartoma, undifferentiated embryonal sarcoma
- Biliary: cyst, rhabdomyosarcoma
- Infectious
- Hematoma
- Pseudomasses: focal fatty infiltration, focal fatty sparing
HEPATIC MASSES - SONOGRAPHY

- Screening technique of choice
- Readily available, fast, less expensive
- Reveals location, extent, solid or cystic nature, and assess vascular invasion
- If normal, no further imaging
- If US not optimal or shows a mass → CT or MRI
<table>
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<th>Age</th>
<th>Newborn</th>
<th>&lt;3 years</th>
<th>3-10 years</th>
<th>&gt;10 years</th>
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ALPHA FETOPROTEIN

- Elevated
  - Hepatoblastoma (80 - 90%)
  - Hepatocellular carcinoma (60 - 90%)
- Normal values in infants < 6m can be higher than in older children
  - particularly important in premature babies

- Normal
  - Hemangioma
  - Hemangioendothelioma
  - Sarcoma
  - Metastases
  - Fibrolamellar HCC
HEPATIC TUMORS IN CHILDREN

- Metastatic liver lesions in children
  - Neuroblastoma
  - Wilms
  - Lymphoma
Newborn 0d with left adrenal neuroblastoma
6y : lymphoma
HEPATOBLASTOMA

- Most common malignant primary pediatric liver tumor (75%)
- Typically 6m-3y; 90% <5y; 4% at birth
- 5% associated with Beckwith-Wiedemann, familial adenomatous polyposis, Gardner syndromes
- Association with low-birth weight (relative risk in <1500g = 15.64 c/w >2500g)
HEPATOBLASTOMA - IMAGING

- Appearances are not specific
- Any liver mass in a child should be considered hepatoblastoma unless specific clinical or imaging features suggest otherwise
HEPATOCELLULAR CARCINOMA

- Older age group than hepatoblastoma
- Peak at 12-15y; rarely before 4y
- In ~50%, preexisting liver disease (hepatitis B, tyrosinemia, type I glycogen storage disease, biliary atresia, familial cholestasis, hyperalimentation, Wilson disease)
HEPATOCELLULAR CARCINOMA

- Imaging features are not specific
15y : hepatitis B
11y : tyrosinemia
FOCAL NODULAR HYPERPLASIA

- Regenerative nodule possibly in response to vascular malformation and/or vascular injury
- May present at any age (youngest in literature 43 days old)
- Often incidental
FOCAL NODULAR HYPERPLASIA

- Abnormal nodular architecture
- Malformed vessels
- Cholangiolar proliferation
- Composed of nodules surrounded by radiating fibrous septa originating from a central scar
- Centrifugal arterial flow
FOCAL NODULAR HYPERPLASIA

- Often not well seen
- Isoechoic or slightly hypo- or hyperechoic
- Mass effect on adjacent vessels
- Deformity of liver contours
- Hypoechoic halo: compressed parenchyma
- Spoke-wheel arterial pattern
10y : incidental finding
6y : back pain, resolved at time of US
FOCAL NODULAR HYPERPLASIA

- Multiple lesions in 20-25% of FNH (adults)
- Often lack central scar
- Becoming more common in oncology patients after completion of therapy
10y : right adrenal neuroblastoma off-therapy for 4 years
HEPATIC HEMANGIOMAS

Terminology is confusing:

- Hemangioma of infancy (new term for lesions previously known as hemangioendothelioma type I & II or capillary hemangioma)
- Congenital hemangiomas (RICH described in liver)
- Hemangioendothelioma (specific types, intermediate aggressiveness; in children: kaposiform & epithelioid types)
- Cavernous hemangioma – adult type hemangioma (most are venous malformations)
HEMANGIOMA OF INFANCY

- Neoplasm of endothelial cells
- Similar to infantile hemangiomas in skin and other soft tissues
- Characteristic phases of rapid growth and spontaneous involution
- Histology: GLUT 1 (+)
HEMANGIOMA OF INFANCY

- In liver, often multifocal
- May be associated with hemangiomas elsewhere
- Usually incidental although may present with CHF
HEMANGIOMA OF INFANCY

- Usually well defined margins, hypoechoic or heterogeneous, less common hyperechoic
- Some associated with signs of high flow and may have A-V shunting
1y: incidental finding
4m: CT done for presumed primary pulmonary hypertension showed liver abnormality
FOCAL HEMANGIOMA

- Most are equivalent to RICH
- Peak of growth at birth followed by rapid involution
- Histology: GLUT 1 (-)
- US: solitary lesion, hypervascular, more heterogeneous
incidental finding during work-up for 2-vessel cord: RICH
RICH: Follow-up at 5m of age
MESENCHYMAL HAMARTOMA

- Benign developmental malformation
- Mixture of bile ducts, hepatocytes, vessels, and mesenchyma without lymphatics
- Mainly cystic (spaces 5 mm - 15 cm) due to fluid accumulation although with variable solid component (stroma)
- 85% in < 2y
- Enlarging abdomen but o/w asymptomatic
- Lab usually normal
MESENCHYMAL HAMARTOMA

- Multilocular mass containing cystic spaces with anechoic or hypoechoic fluid separated by echogenic septa
- If cystic spaces are small → may appear solid
3m : hepatomegaly
UNDIFFERENTIATED EMBRYONAL SARCOMA

- Rare, mainly in older children and adolescents
- Fever, painful mass, normal αFP
- Large mass 7-20 cm
- Undifferentiated sarcomatous tissue in a myxoid matrix (solid-cystic) → US appearance
- Poor prognosis
12y : hepatomegaly
US is extremely useful and often only imaging modality required
Neonatal hepatitis and biliary atresia are common causes.
Differentiation is crucial for management:
- Surgical for biliary atresia
- Medical in hepatitis
NEONATAL HEPATITIS

- Nonspecific inflammation:
  + Infection (serum hepatitis, CMV, herpes simplex, toxoplasmosis, rubella, protozoa, syphilis)
  + Defects of metabolism (α1-antitrypsin deficiency, galactosemia, GSD, tyrosinosis)
  + Familial cholestatic syndromes

- US findings are nonspecific and not always present
  + Hepatomegaly
  + Hyperechoic parenchyma
BILIARY ATRESIA

- Congenital obliterative cholangiopathy of unknown etiology
- Affects both intra- and extrahepatic ducts
- Types:
  - Non-syndromic
  - Syndromic (heterotaxy, interrupted IVC, preduodenal portal vein, intestinal malrotation, cardiac defects, polysplenia)
- Early diagnosis and prompt surgery crucial for prognosis
BILIARY ATRESIA

- US is useful in diagnosis & highly accurate in experienced hands
- Some authors show high sensitivity and specificity of US suggesting that the combination of US findings may obviate biopsy and indicate surgery
- NASPGHAN guidelines recommend that US cannot be used to rule out BA
- Scintigraphy with hepatobiliary agents, cholangiography and liver biopsy may also be needed
BILIARY ATRESIA: US DIAGNOSIS

- Gallbladder
  - Small size <19mm or not seen
  - Abnormal shape
  - Wall irregularity
- Triangular cord sign
- Non-visualization of CBD
- Hepatomegaly
- Splenomegaly
- Prominent hepatic artery (2.2mm  0.59)
BILIARY ATRESIA: US DIAGNOSIS

- Gallbladder:
  - Seen 77%; not seen 23%
  - <19mm 61%; ≥19mm 39%
  - Abnormal shape 70%; normal shape 30%
  - Wall irregularity 91%; normal wall 9%

- Triangular cord sign:
  - Present 73%; absent 27% (false + 0.7%)

- CBD:
  - Not detected 93%; detected 7%

Humphrey TM & Stringer MD. Radiology 2007;244:845-851
BILIARY ATRESIA

2m: conjugated hyperbilirubinemia & hepatosplenomegaly
CHOLEDODCHAL CYST

- Congenital dilatation of the common bile duct
- Diagnosis: <1y 30%, 1y-10y 50%; >10y 20%
- Classic triad: jaundice, abdominal pain and mass only seen in 20-50%
- Etiology unknown
CHOLEDOCHAL CYST

- Well defined, fluid-filled mass in or near porta hepatis, separate from the gallbladder
- Intrahepatic biliary dilatation in 50%, limited to central hepatic ducts
- Complications: rupture, lithiasis (cyst, gallbladder, pancreatic duct), neoplasia, pancreatitis
TODANI CLASSIFICATION (1977)

- Type I: Segmental or diffuse fusiform dilatation of CBD (75-95%)
- Type II: Diverticulum
- Type III: Choledochocoele
- Type IV: Multiple extrahepatic cysts (B) or associated with Caroli-type intrahepatic biliary cysts (A)
- Type V: Caroli disease

Nievelstein et al. Pediatr Radiol 2011; 41:55–75
CHOLEDOCHAL CYST

4w : antenatal diagnosis of RUQ cyst
10m : vomiting
CAROLI DISEASE/SYNDROME

- Autosomal recessive congenital cystic dilatation of intrahepatic ducts
- Termed Caroli syndrome when associated with congenital hepatic fibrosis, more common in children and when symptomatic is due to portal hypertension
- Caroli disease in adults presents clinically with calculus formation & recurrent bacterial cholangitis
- Associated with renal tubular ectasia → ARPKD
CAROLI DISEASE/SYNDROME

- Multiple dilated tubular and cystic structures
- Clustered or diffuse
- Echogenic protrusions or bands that extend into lumen of dilated duct → “echogenic dot” sign
LANGERHANS CELL HISTIOCYTOSIS (LCH)

- Rare, peak 1-3 years
- Proliferation of antigen-presenting dendritic cells (Langerhans cells) may be reaction to infection, cytokines or a clonal proliferative disorder
- Liver involvement: 10% at presentation and >14% over course of disease
- Liver LCH involvement of biliary ducts → sclerosing cholangitis → biliary cirrhosis → liver failure
- Liver damage may progress despite LCH regression
LANGERHANS CELL HISTIOCYTOSIS (LCH)

- Echogenic wall thickening of intrahepatic biliary ducts (periportal distribution)
- May result in focal biliary dilatation