Imaging of Pediatric Neoplastic Liver Masses

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Presentation of Liver Masses

- Signs & Symptoms
  - Pain, anorexia, weight loss, fever, jaundice, congestive heart failure
- Physical Exam
  - Palpable liver mass
  - Abdominal distension

Imaging??

- US
  - Frequently the first modality: available & fast
  - Pt must fast 4-6 hours prior to procedure
  - Determines the location & extent of mass
  - Solid vs cystic
- CT
  - Use when US information isn't sufficient
  - Sedation of young children necessary
  - Dual-phase

Pediatric Hepatic Tumors

- Benign
  - Hemangioendothelioma
  - Mesenchymal hamartoma
  - Cavernous hemangioma
  - Focal nodular hyperplasia
  - Hepatic adenoma
- Malignant
  - Hepatoblastoma
  - Hepatocellular carcinoma
  - Posttransplant Lymphoproliferative Disorder
  - Metastatic disease
  - Embryonal sarcoma
  - Primary endodermal sinus tumor

The Benign Neoplasms

Hemangioendothelioma

Mesenchymal hamartoma

Cavernous hemangioma

Focal nodular hyperplasia

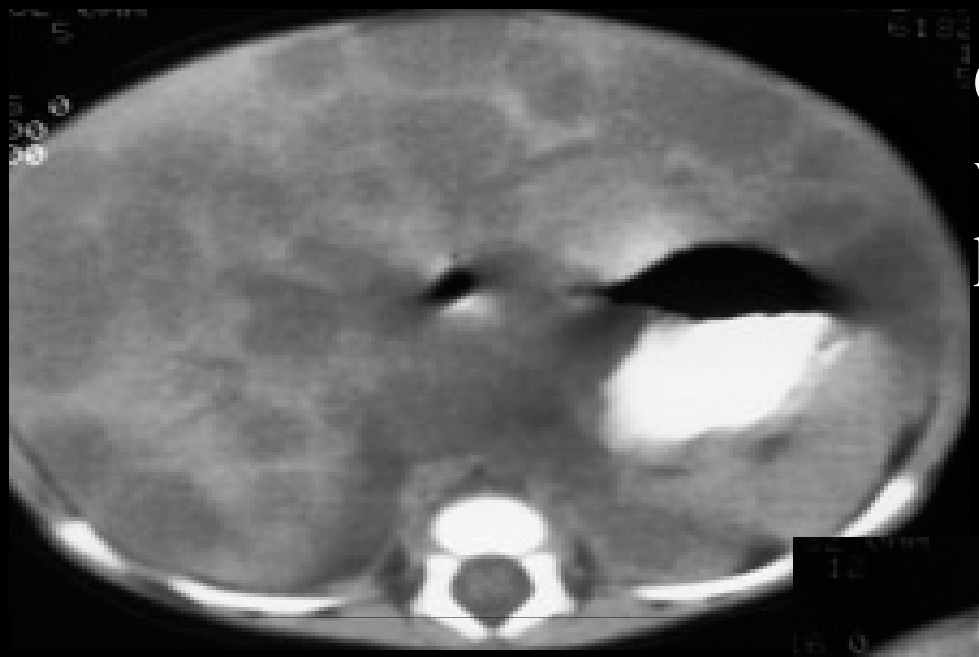
Hepatic adenoma

Hemangioendothelioma

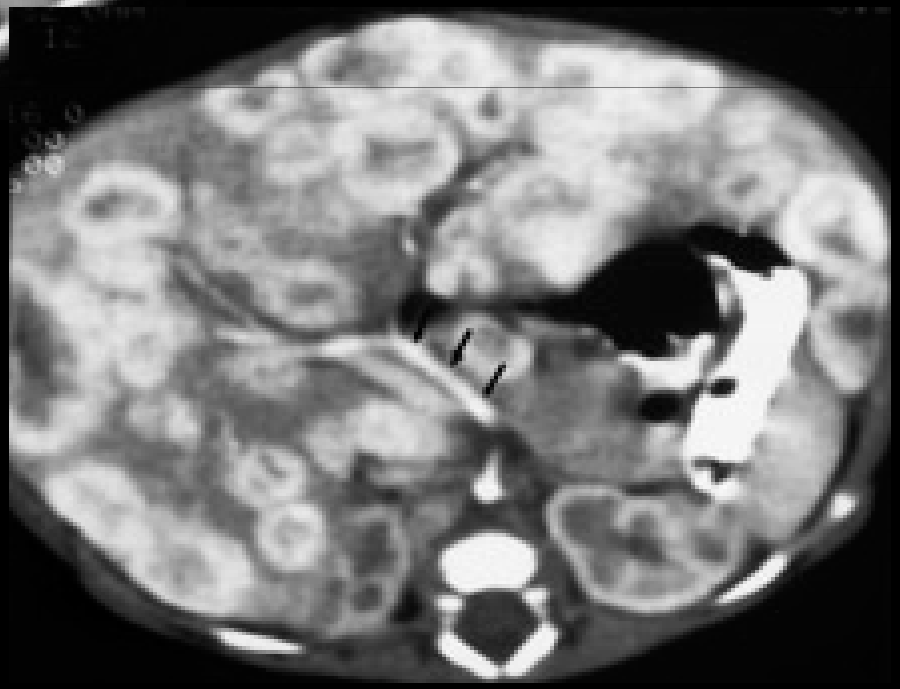
- Most common pediatric benign hepatic tumor
- Patients < 6 months old
- Presenting signs and symptoms
  - CHF
  - Hepatomegaly
  - Thrombocytopenia
  - Hemoperitoneum

Hemangioendothelioma: Characteristics

- Gross Appearance:
  - Multiple nodules
  - Solitary or multifocal
- Histologically:
  - Vascular tumor of mesenchyme
  - Variable areas of fibrosis, calcification, hemorrhage, & cystic degeneration
- US
  - Variable
- Unenhanced CT
  - Low attenuation
  - 50% w/ calcifications
- Post IV contrast CT
  - Large tumors: peripheral puddling of contrast
  - Small tumors: immediate enhancement
  - Variable intensity due to fibrosis, necrosis, & hemorrhage
  - Descending aorta distal to celiac artery appears small due to shunting of CO towards the tumor



CT of 1-month-old girl with multiple hemangioendotheliomas



•Above: Precontrast CT of heterogeneous liver

•Right: CT arterial-phase. Enhancement of lesions. Arrows demonstrate large hepatic artery.

Hemangioendothelioma: Treatment

- Natural regression within 12-18 months
- Medical management includes:
  - Digitalis
  - Diurectis
  - Steroids
  - Interferon
- Chemotherapy, irradiation, embolization, and/or surgery are considered if medical management fails

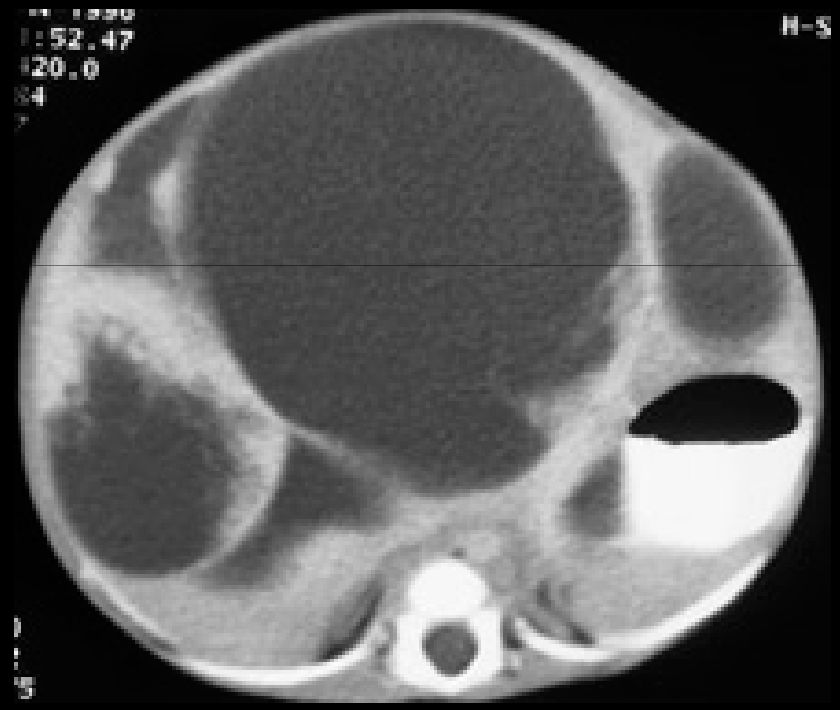
Mesenchymal Hamartoma

- Benign tumor arising from portal mesenchyme, likely a congenital malformation
- Slightly increased prevalence in males
- 2-3 years of age
- Presenting signs and symptoms
  - Asymptomatic abdominal mass or distension
  - CHF and ascites are uncommon on presentation
- Treatment
  - Surgical resection

Mesenchymal Hamartoma: Characteristics

- Gross Appearance:
  - Well-circumscribed
  - Frequently right lobe
- Pathologic Appearance:
  - Multiple fluid-filled cysts separated by fibrous stroma with mesenchyme, abnormal bile ducts, & hepatocytes
- US
  - Anechoic or hypoechoic
- CT
  - Low attenuation
  - Appearance variable due to protein in cystic fluid
  - Stromal enhancement with contrast

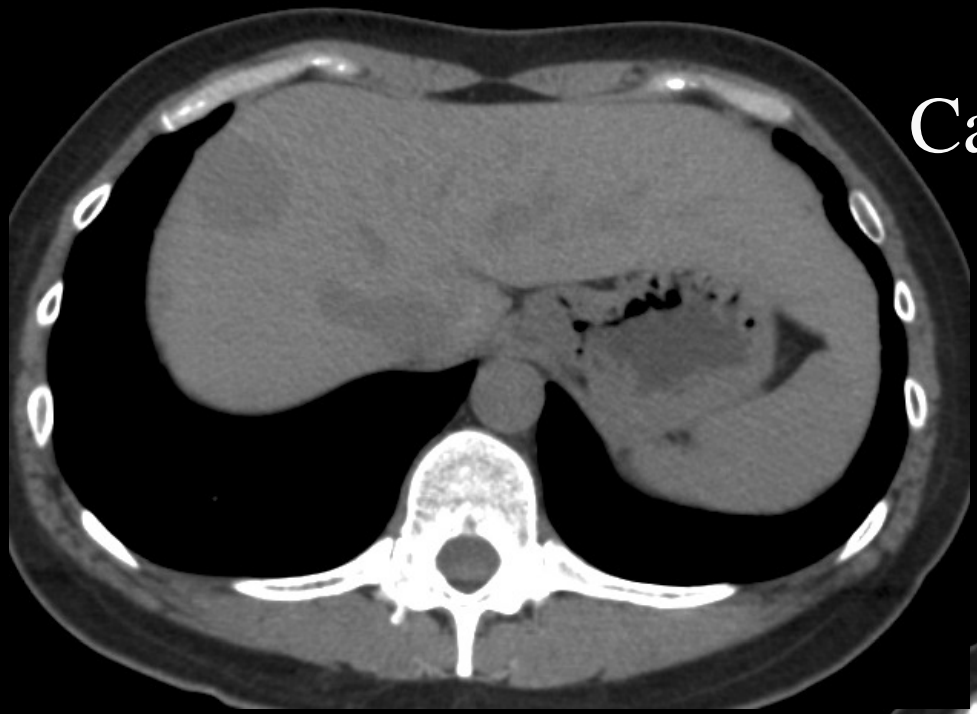
Mesenchymal Hamartoma. Contrast-enhanced CT.

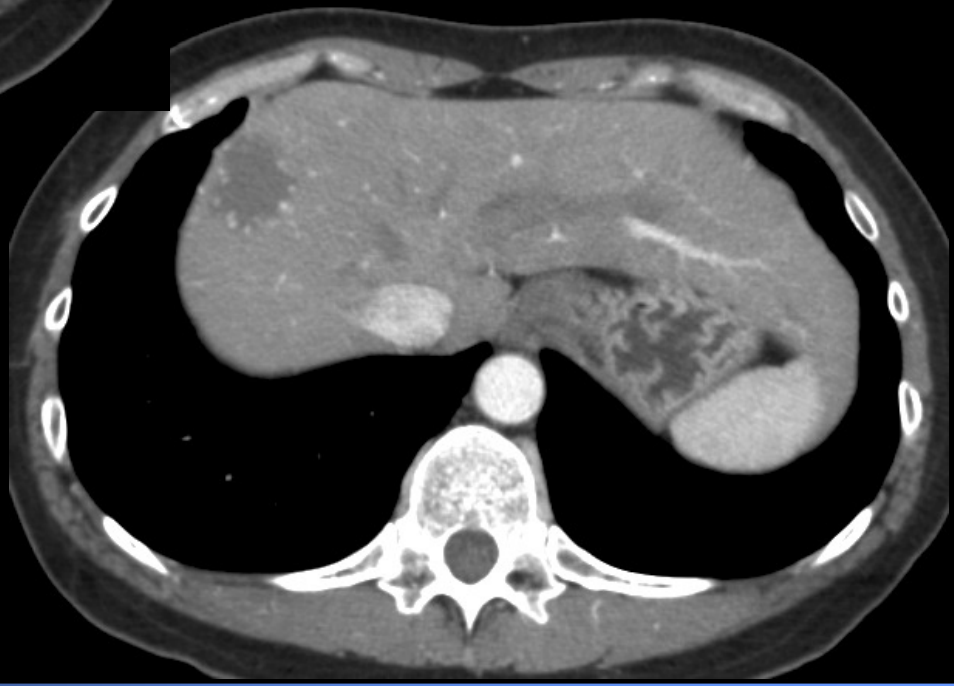


Cavernous Hemangioma

- Older children & adolescents
- Small, solitary, assymptomatic, & an incidental finding
- Posterior segment of right lobe
- No malignant potential
- Pathologically:
  - Multiple blood filled spaces lined with mature endothelial cells
- US
  - Hyperechoic
  - Homogeneous
  - Well-defined
- CT
  - Nonenhanced: hypoattenuating
  - Arterial-phase: nodular peripheral enhancement
  - Similar to hemangioendotheliomas

Cavernous Hemangioma

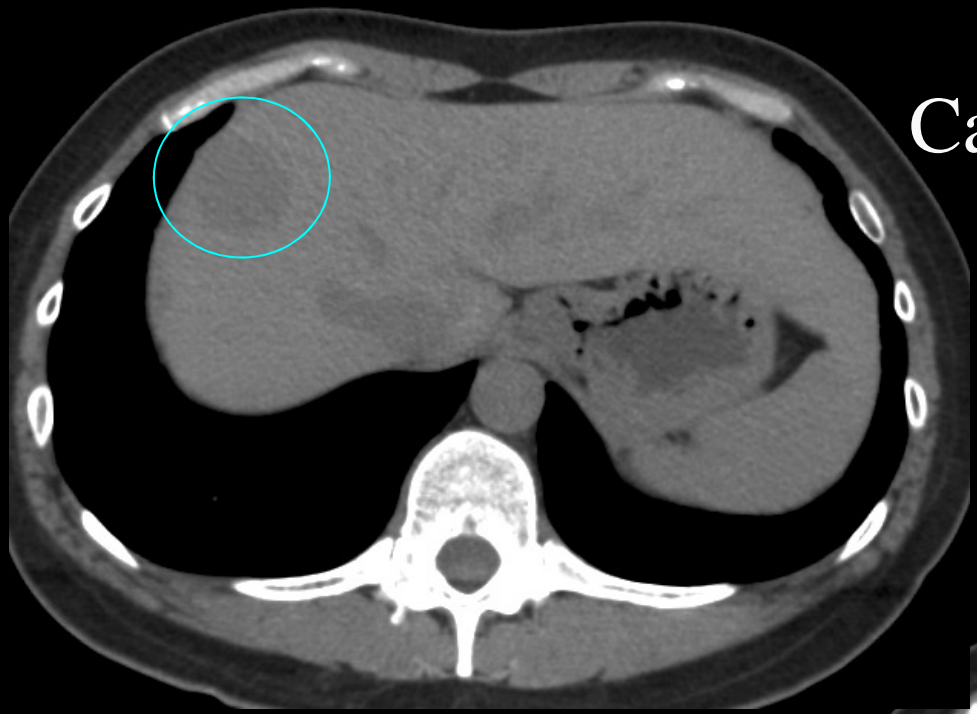


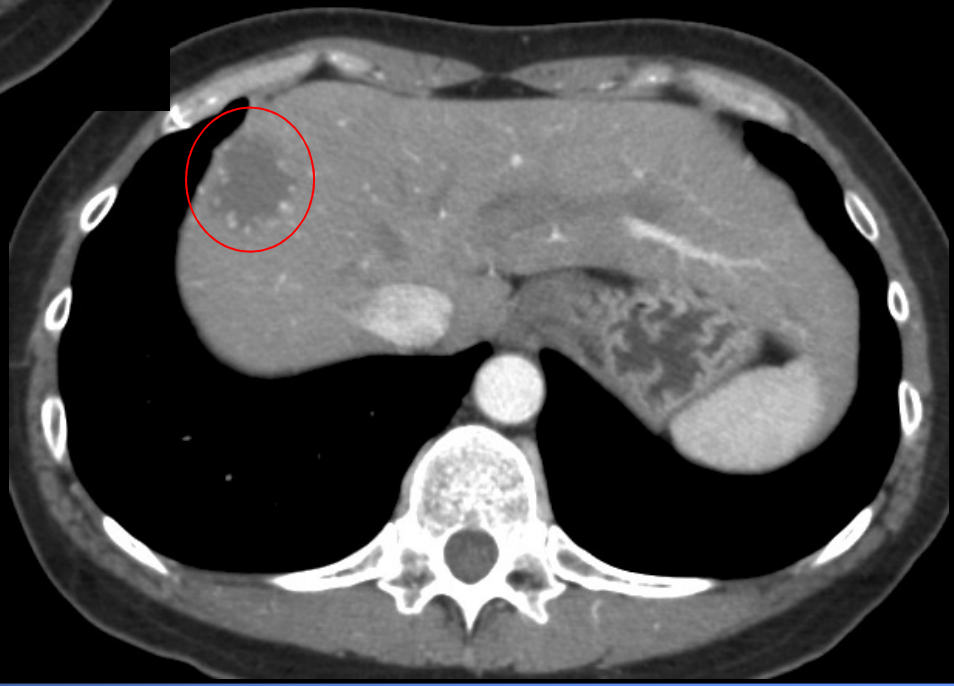


Above: Non-contrast CT displaying hypoattenuating mass.

Right: Arterial-phase CT demonstrating nodular peripheral enhancement of the mass.

Cavernous Hemangioma





Above: Non-contrast CT displaying hypoattenuating mass.

Right: Arterial-phase CT demonstrating nodular peripheral enhancement of the mass.

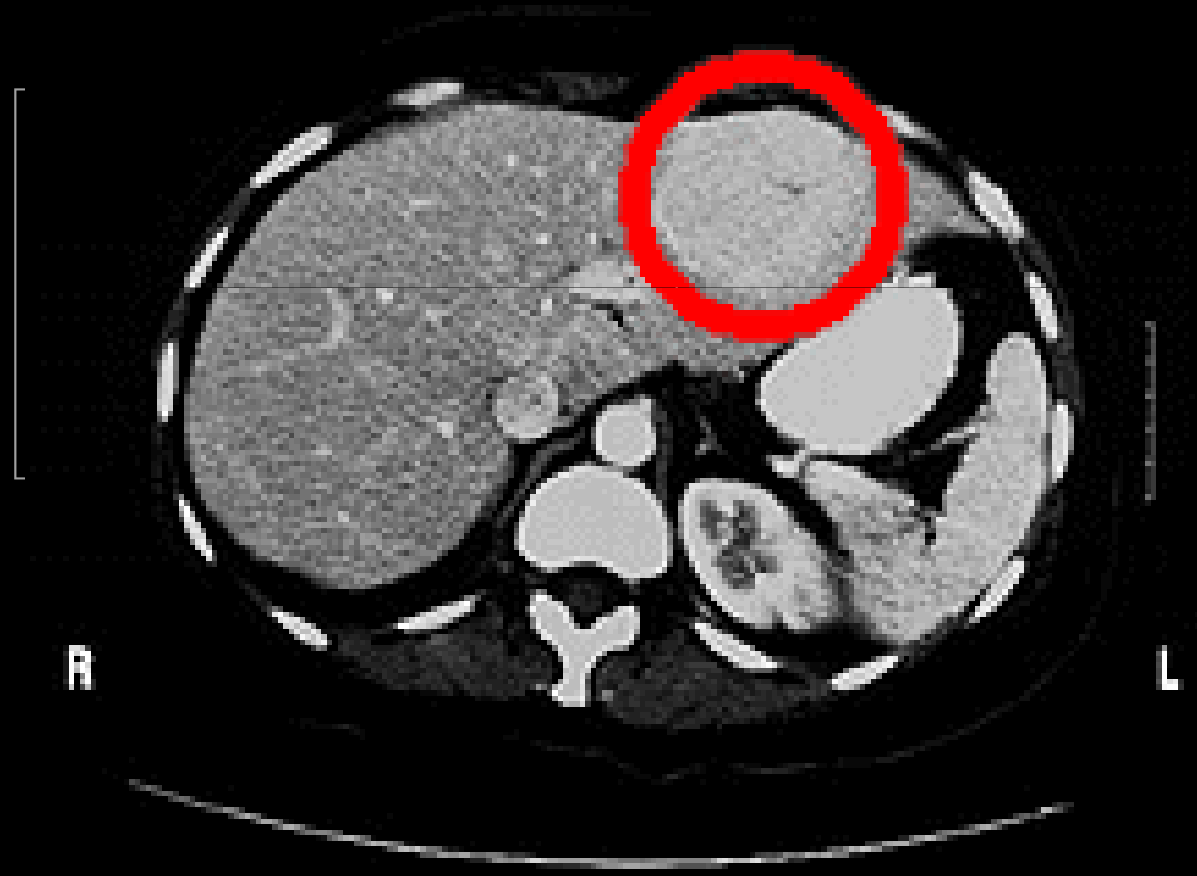
Focal Nodular Hyperplasia

- < 5% of pediatric hepatic masses
- Histological
  - Normal hepatocytes, bile ducts, & Kupffer cells
- Near liver surface
- Central stellate scar with bile ducts & arteries
- US
  - Well defined
  - homogeneous

CT

- Hypo/isoattenuating relative to liver
- Contrast enhanced CT demonstrates diffuse enhancement with rapid washout
- Central scar will remain enhanced due to delayed wash out
- <5% have small calcifications

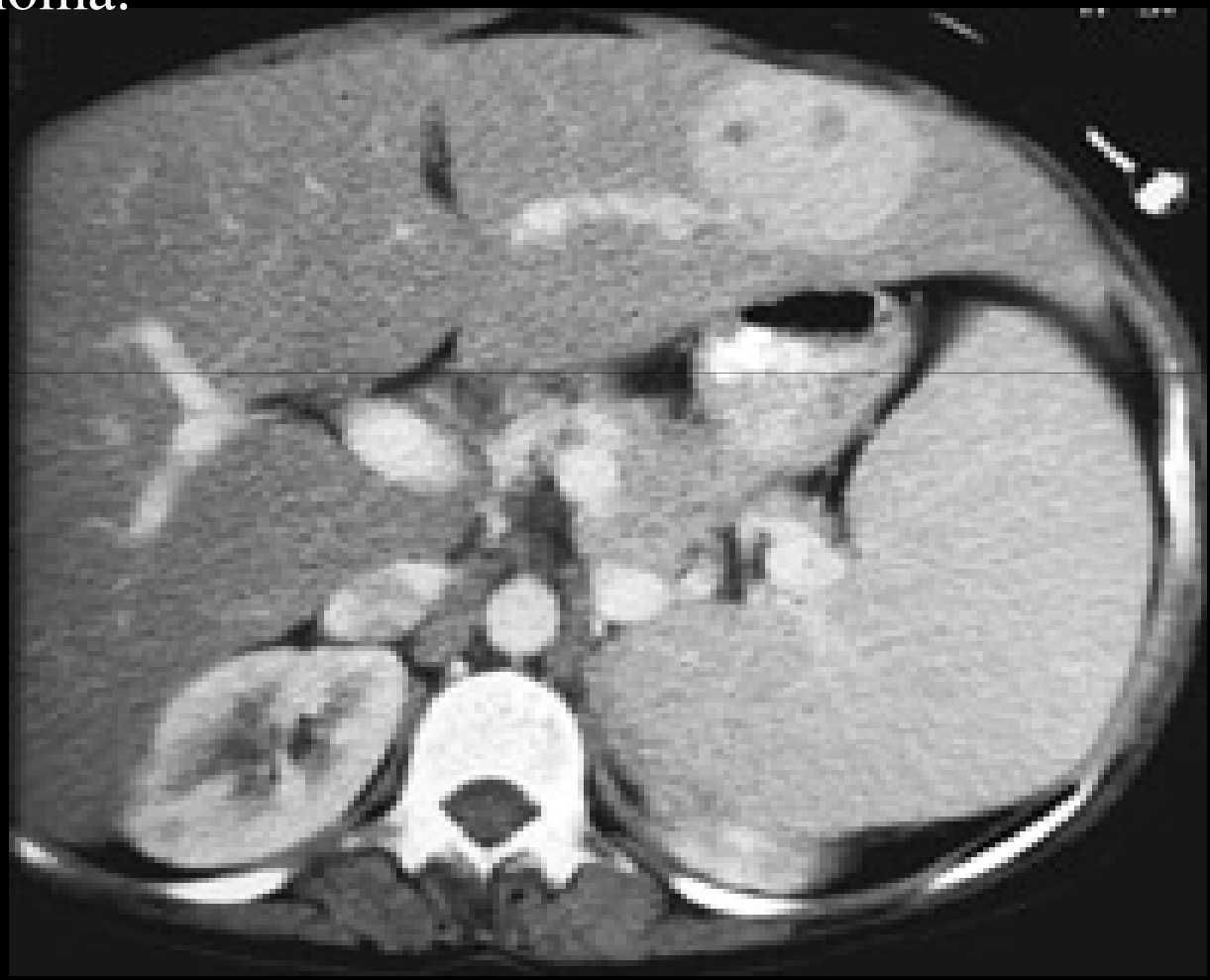
Focal Nodular Hyperplasia



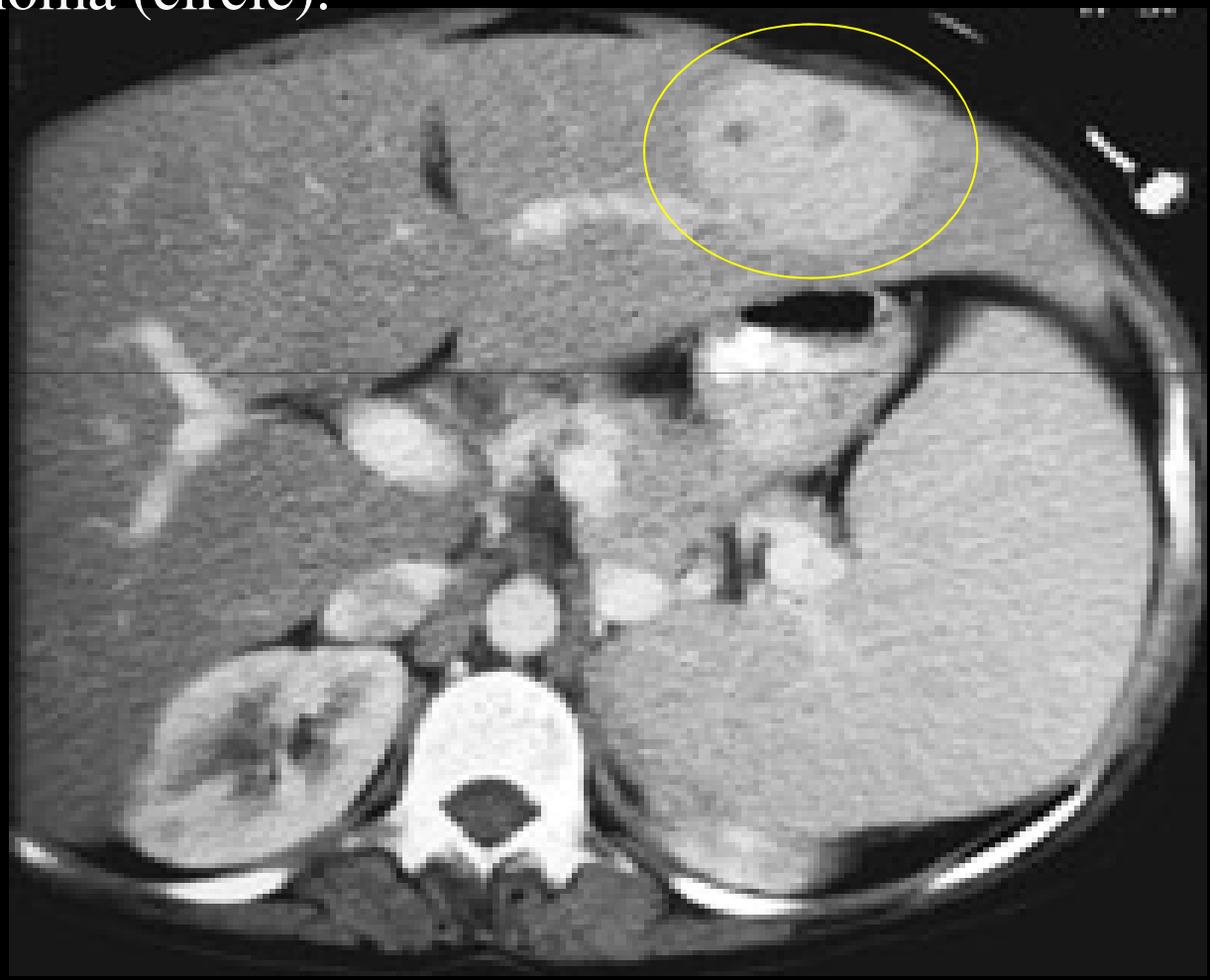
Hepatic Adenomas

- <5% of childhood hepatic tumors
  - Associations:
    - von Gierke's disease
    - Fanconi's anemia
    - Galactosemia
    - Anabolic Steroids
- Histologically:
  - Normal hepatocytes
  - No bile ducts or portal tracts
- US
  - Variable echogenicity
- CT
  - Unenhanced: hypoattenuating
  - Early enhancement with contrast
  - Heterogenous due to hemorrhage, necrosis, glycogen, or fat

CT scan with contrast, arterial-phase. 12yo female with von Gierke's. Enhancing heterogeneous adenoma.



CT scan with contrast, arterial-phase. 12yo female with von Gierke's. Enhancing heterogeneous adenoma (circle).



The Malignant Neoplasms

Hepatoblastoma

Hepatocellular carcinoma

Posttransplant Lymphoproliferative Disorder

Metastatic disease

Embryonal sarcoma

Primary endodermal sinus tumor

Hepatoblastoma

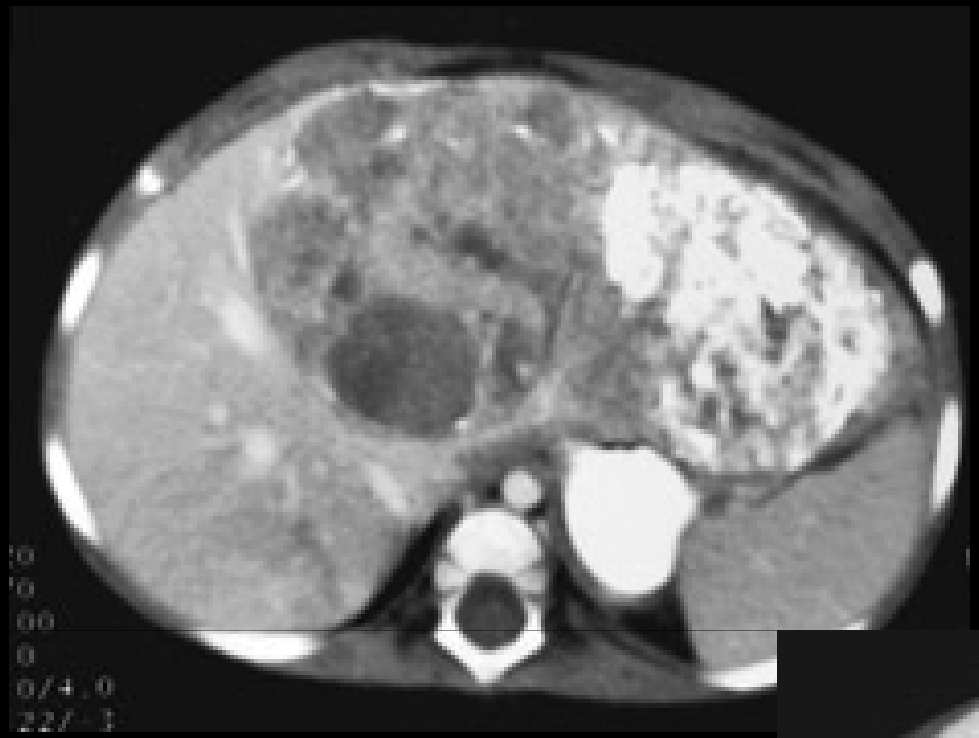
- 45% of pediatric liver masses
- Infants & young kids
- Associations:
  - Beckwith-Wiedemann
  - Hemihypertrophy
  - FAS
  - Familial Polyposis Coli
  - Gardner's Syndrome
- 65% 2-yr survival

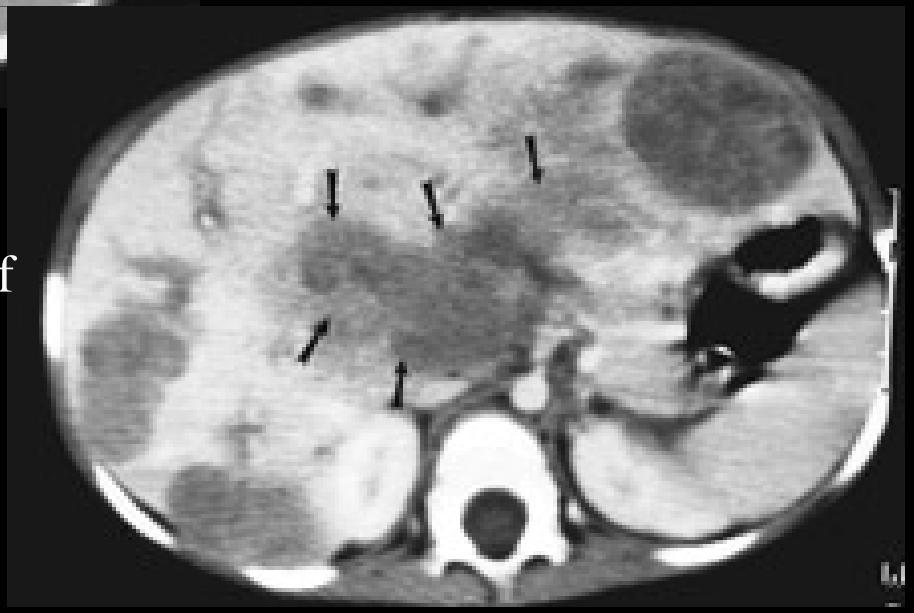
- Presenting Signs & Symptoms
  - Usually asymptomatic
  - Abdominal pain
  - Anorexia
  - Weight loss
  - Jaundice
  - Precocious puberty
  - Osteopenia
  - Elevated serum AFP

Hepatoblastoma: Characteristics

- Unifocal, freq R lobe
- No assoc. cirrhosis
- 10-20% metastasize
  - Lungs, porta hepatis, brain, skeleton
- Histologically
  - Primitive epithelial cells resembling fetal liver
- US
  - Heterogeneous mass w/ distortion of vascular parenchymal & calcifications
- CT
  - Heterogenous, low attenuation mass
  - Arterial phase: enhancing
  - Venous phase: hypoattenuating
- Imaging may demonstrate
  - Spread to portal lymph nodes
  - Intravascular extension
  - Tumor thrombus

Hepatoblastoma

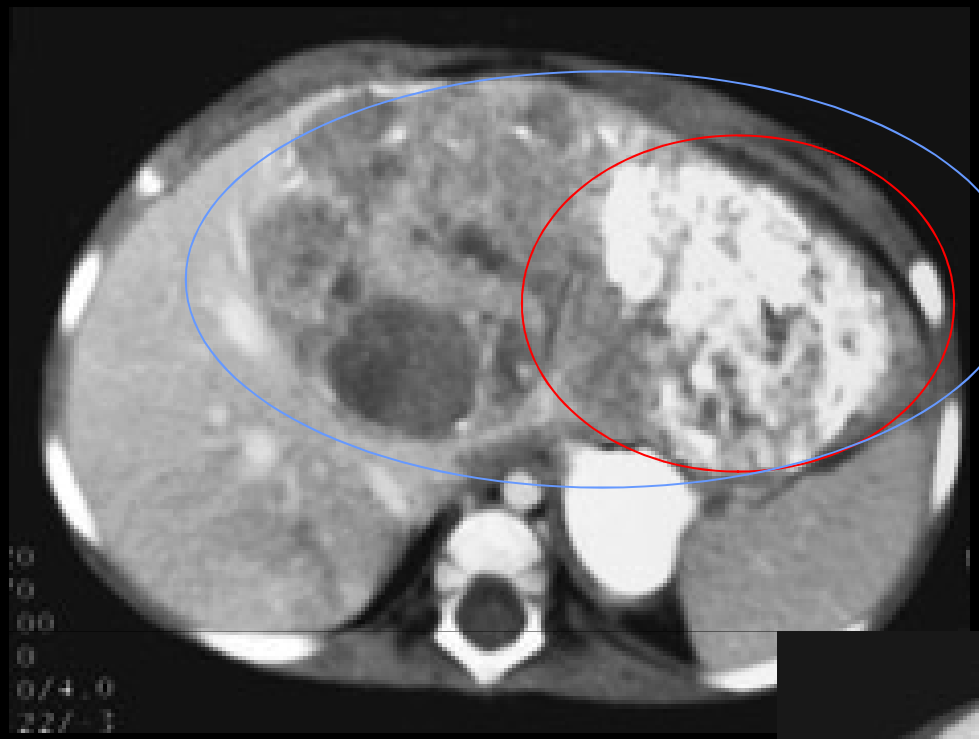


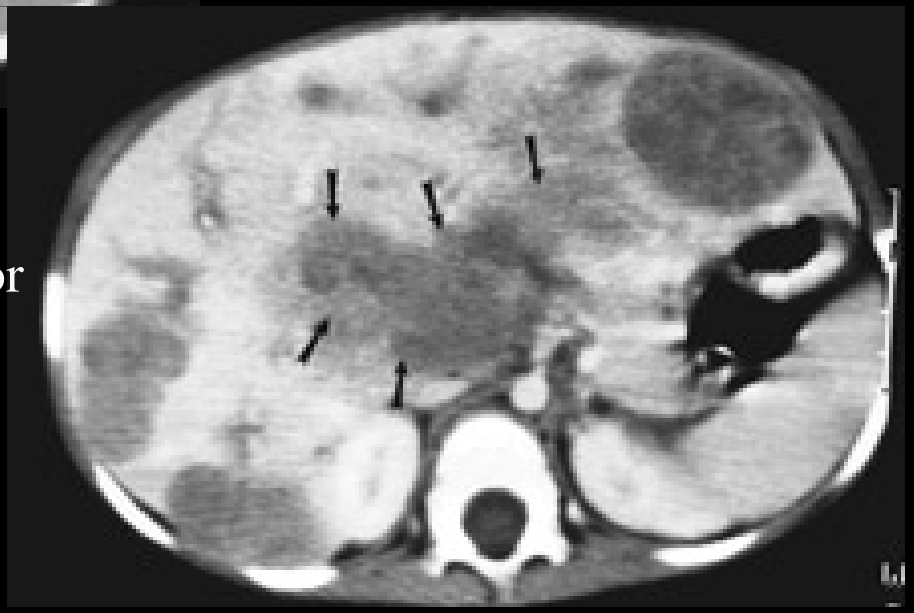


Above: CT 1yo girl. Large, highly calcified mass of anterior segment of R lobe and entire L lobe.

Right: Multiple low attenuation nodules. Tumor thrombus in main portal veins (arrows).

Hepatoblastoma





Above: CT 1yo girl. Large, highly calcified (red) mass (blue) of anterior segment of R lobe and entire L lobe.

Right: Multiple low attenuation nodules. Tumor thrombus in main portal veins (arrows).

Hepatocellular Carcinoma (HCC)

- 2nd most prevalent pediatric liver malignancy
- Median age: 12yo (range: 5-15)
- Presenting signs & symptoms
  - Abdominal distension
  - R upper quadrant mass
- 50% have elevated AFP
- Assoc. preexisting liver disease in ½ of cases
  - Hepatitis B
  - Type I glycogen storage disease
  - Tyrosinemia
  - Familial cholestatic cirrhosis
  - Hemochromatosis
  - Alpha 1-antitrypsin deficiency

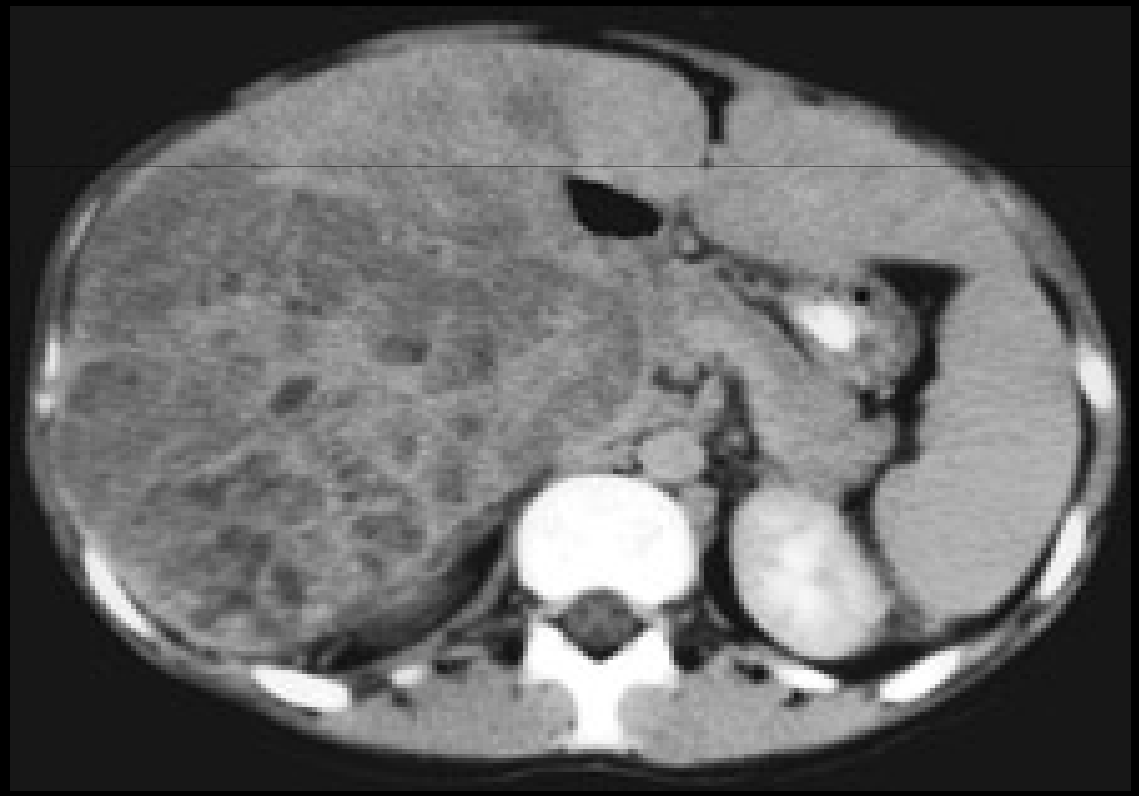
Hepatocellular Carcinoma: Characteristics

- Pathologically
  - Large, pleomorphic multinucleated cells with variable differentiation
- Tumor freq extensive at diagnosis, resectable in <30% of patients
- ≤ 29% survival

- US
  - Heterogeneous, mostly hyperechoic
- CT
  - Low attenuation
  - 25% w/ calcifications
  - Early arterial enhancement with rapid washout

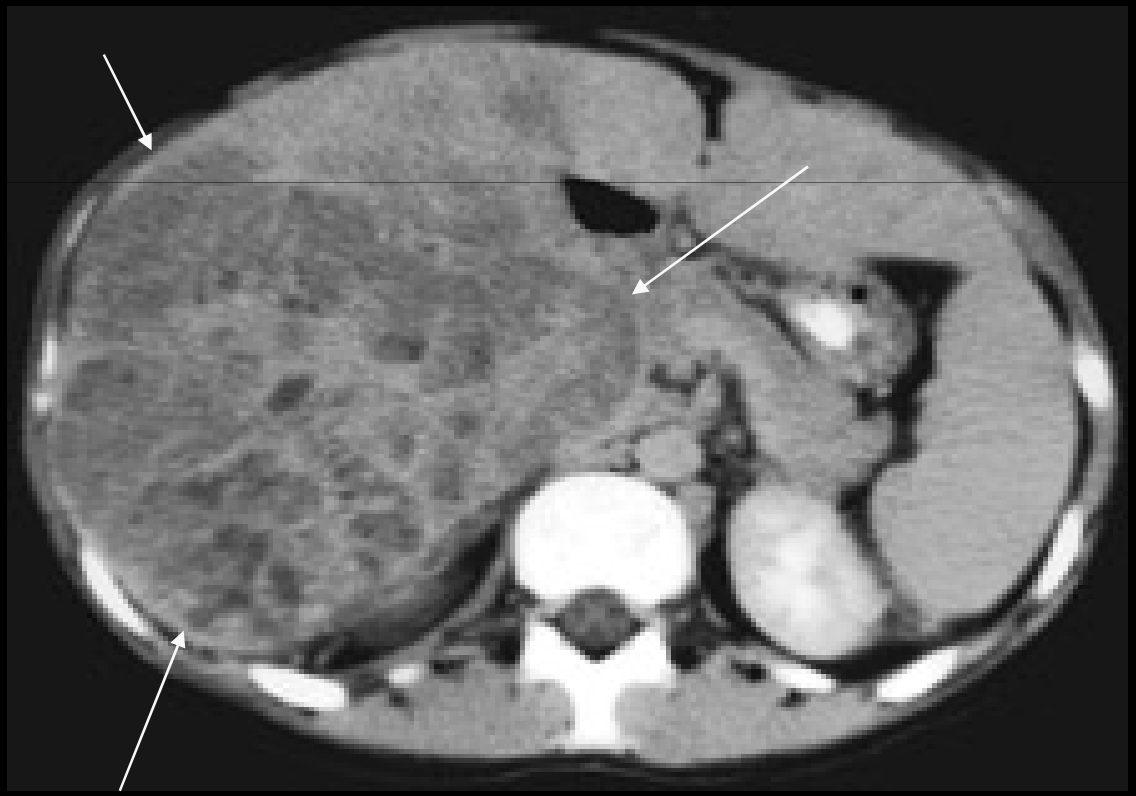
Hepatocellular Carcinoma:

CT w/ contrast. 14yo girl. Large heterogeneous mass of R lobe. Successful surgical outcome as tumor was confined to R lobe.



Hepatocellular Carcinoma:

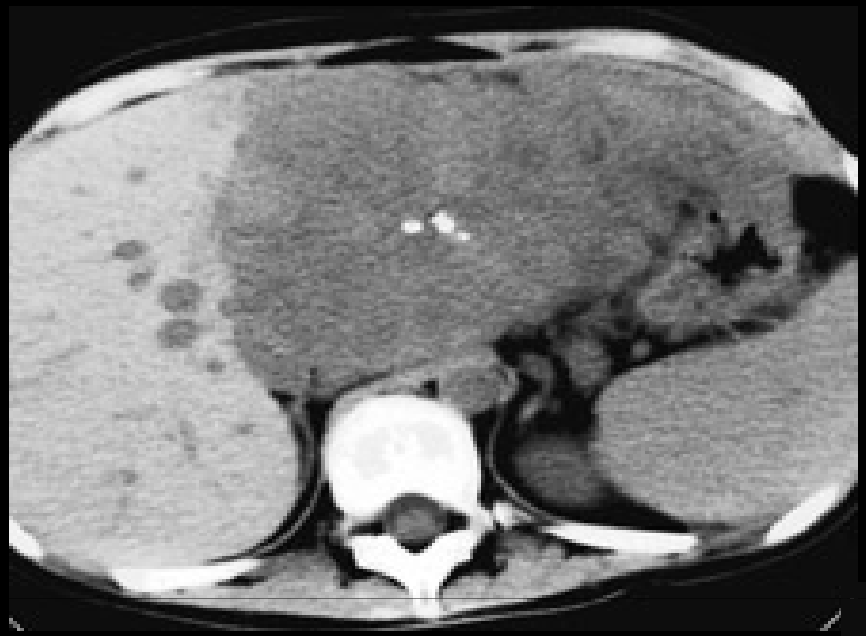
CT w/ contrast. 14yo girl. Large heterogeneous mass (arrows) of R lobe. Successful surgical outcome as confined to R lobe.

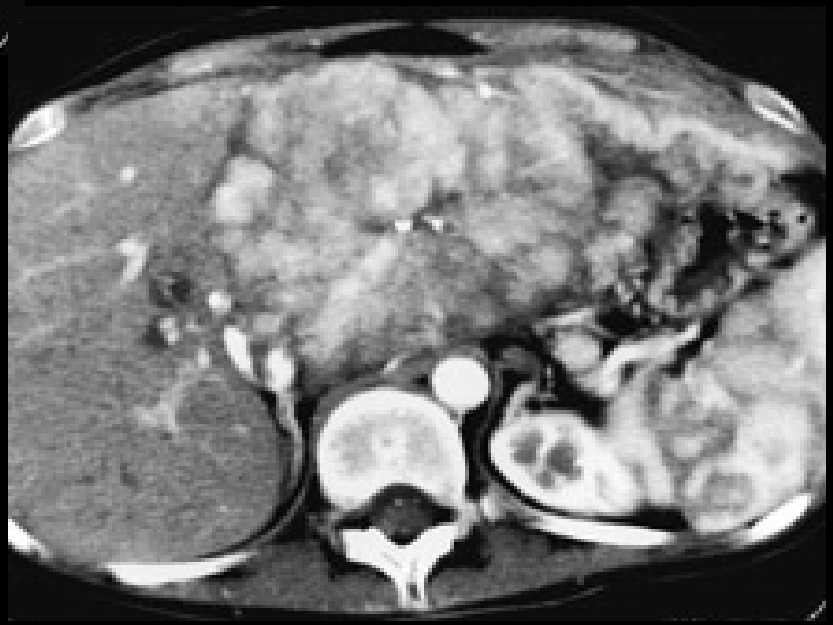


Fibrolamellar Hepatocellular Carcinoma

- Rare subtype of HCC
  - Less aggressive w/ more favorable prognosis
- Adolescents & young adults
- Histologically
  - Eosinophilc hepatocytes separated with thin fibrous bands
- Presentation
  - Hepatomegaly & abdominal pain
  - Normal AFP
- Single, circumscribed mass
- US
  - Mixed echogenicity
- CT
  - Hypoattenuating
  - Contrast enhancing in arterial & venous phases
  - 70% have central scar and calcifications

Fibrolamellar HCC

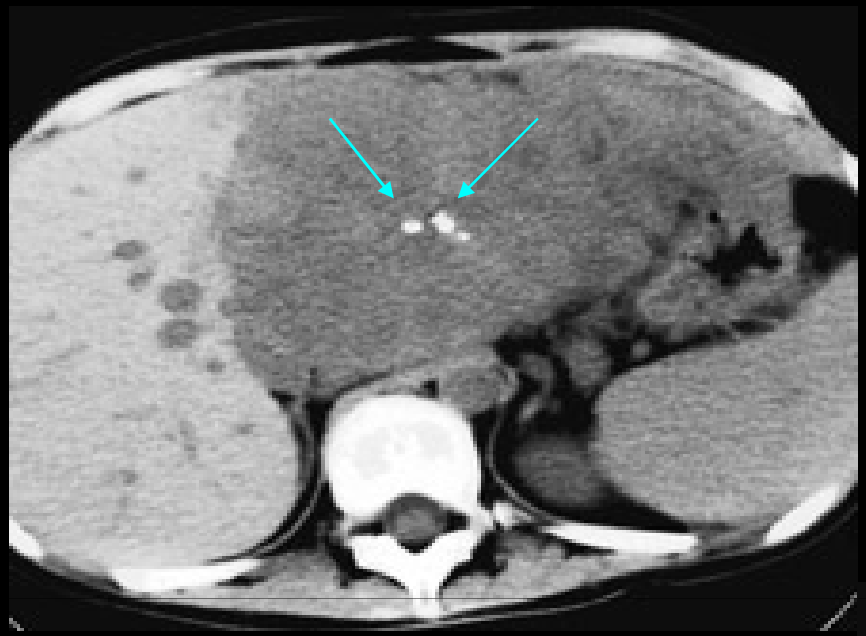


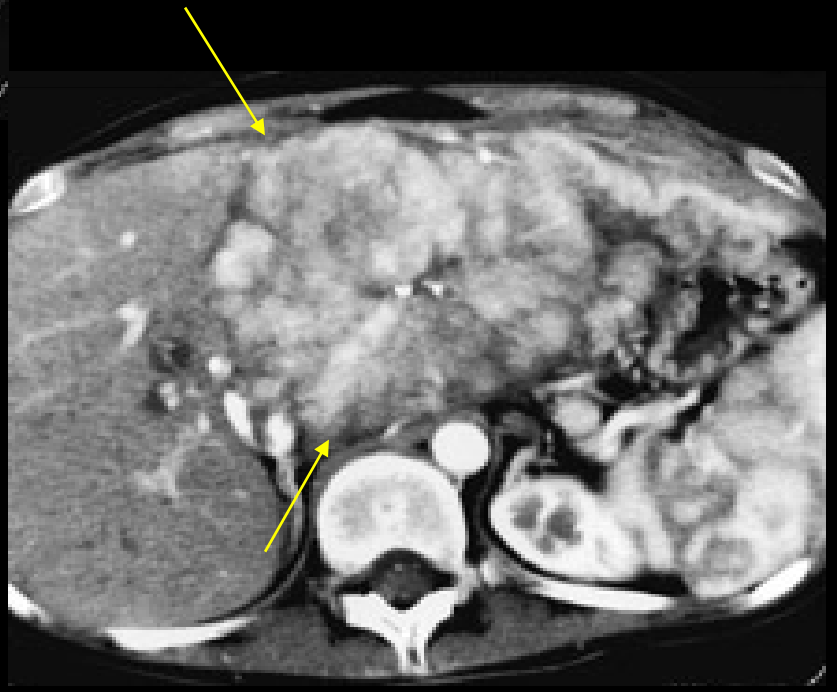


Above: Non-contrast CT displays mass with central calcifications.

Right: Arterial-phase CT displays heterogeneous enhancement of the mass.

Fibrolamellar HCC



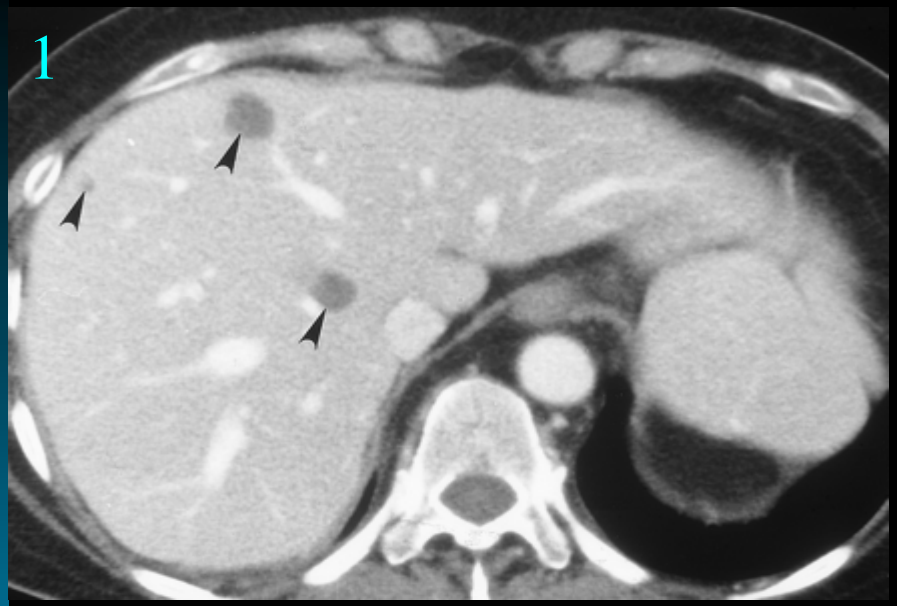


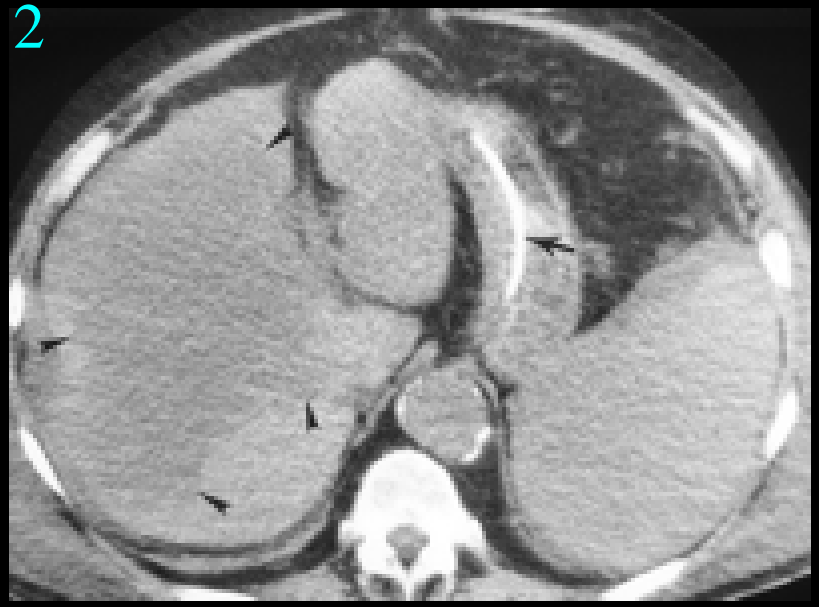
Above: Non-contrast CT displays mass with central calcifications (arrows).

Right: Arterial-phase CT displays heterogeneous enhancement of the mass (arrows).

Posttransplant Lymphoproliferative Disorder (PTLD)

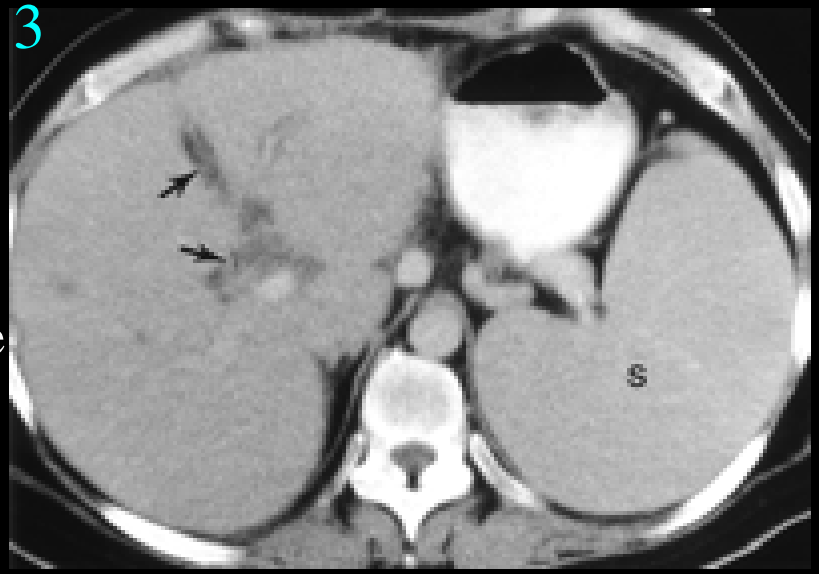
- 50% of kids with PTLD have liver involvement
- Imaging appearance similar to metastases
- Three imaging patterns
  1. Discrete hypoechoic/low-attenuation nodular lesions, 1-4 cm (most common pattern)
  2. Infiltrative, poorly defined lesions, possibly leading to hepatomegaly
  3. Involvement of porta hepatis with frequent periportal infiltration/extension into biliary tree
     - May lead to biliary obstruction





PTLD:

1. CT w/contrast: Well-defined, low-attenuation nodular lesions (arrowheads).
2. CT w/out contrast: Infiltrative, poorly defined lesions (arrowheads). NG tube in stomach (arrow).
3. CT w/contrast: Periportal infiltration (arrows).



Metastatic Disease

- Most common pediatric tumors to metastasize to liver
  - Wilms'
  - Neuroblastoma
  - Rhabdomyosarcoma
  - Lymphoma
  - Leukemia
- Presenting signs & symptoms
  - Hepatomegaly
  - Jaundice
  - Abdominal pain/mass
  - Abnormal LFTs

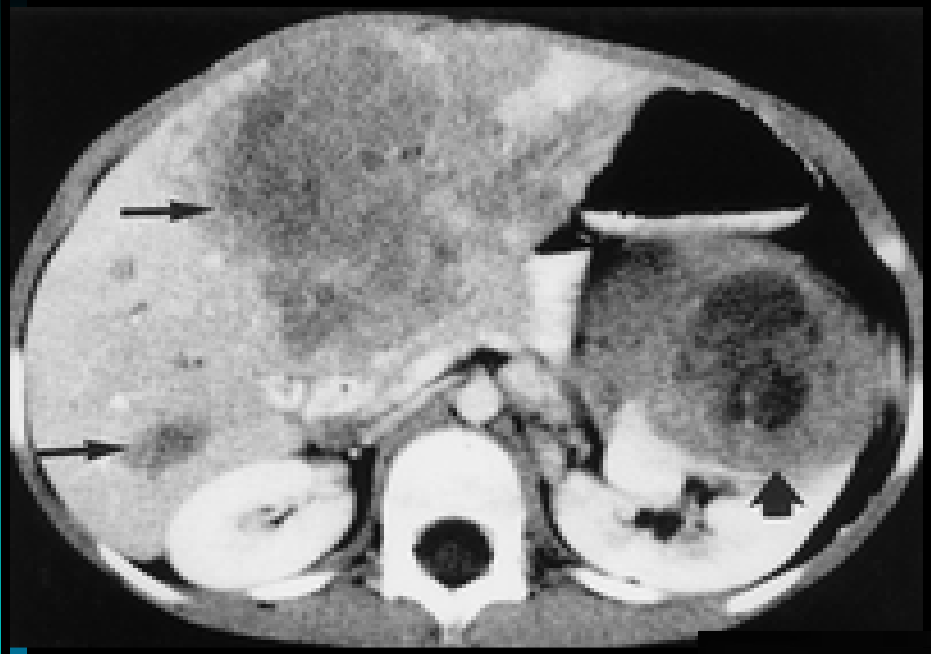
Metastatic Diseases

- Neuroblastoma
  - Stage IV: retroperitoneal mass with metastases to liver/skeleton/nodes
  - Stage IV-S: <1yo small ipsilateral tumors with metastases to liver, skin, & bone marrow, not cortical bone
  - Chemotherapy is primary treatment modality
  - Prognosis dependent on age, stage, and biology
- Wilms' tumor
  - Stage IV: Hematogenous spread to liver via renal vein
  - TXT: Surgery + Chemo + XRT
  - Survival varies with cytological features, 16.7-80.9% 4-yr survival

Metastatic Disease: Characteristics

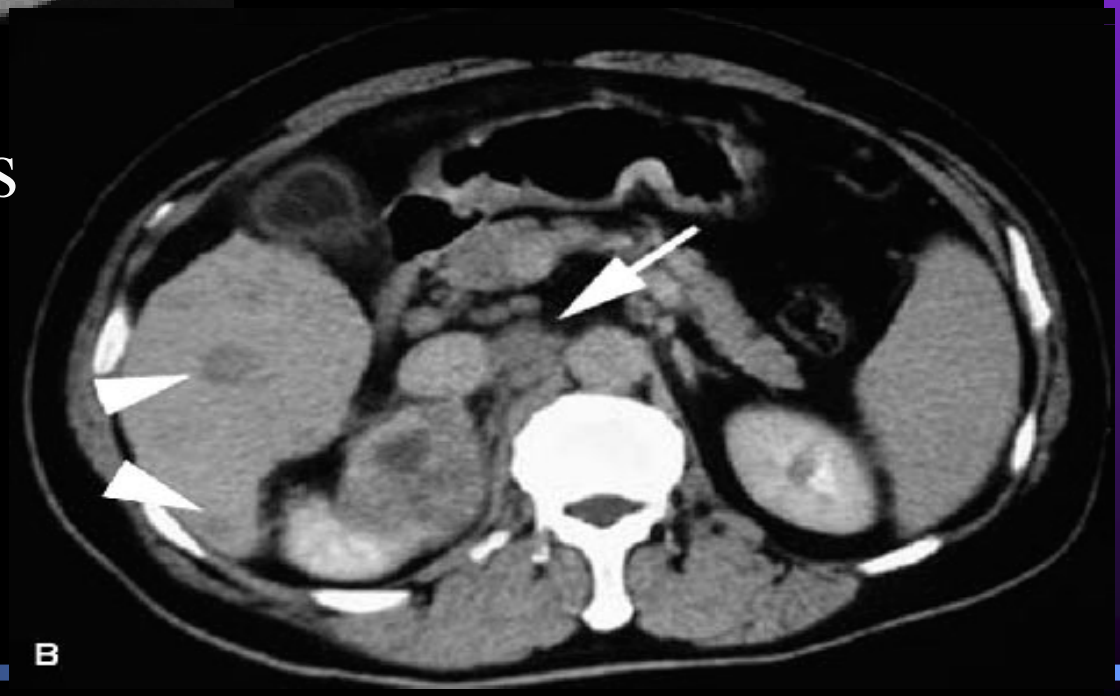
- Neuroblastoma IV-S: diffuse replacement
- All imaging modalities show widespread heterogeneity
- US
  - Hypo/hyperechoic
- CT
  - Contrast: hypoattenuating, w/ occasional peripheral enhancement
  - Hypovascular in arterial and venous phases
  - Possible mass effect w/ displacement of vessels

Hepatic Metastases



Left: CT w/contrast 4yo male. Wilms' tumor (thick arrow) & hepatic metastases (thin arrows).

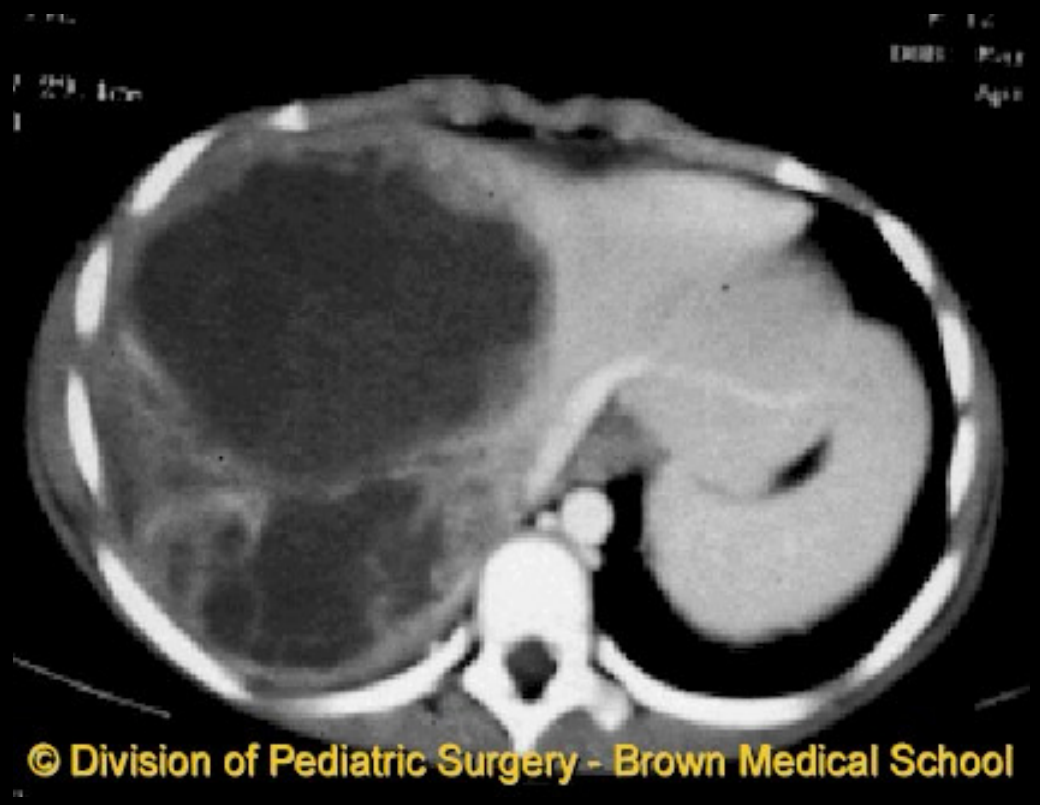
Below: CT. Neuroblastoma hepatic metastases (arrowheads) & large lymph node (arrow).



Embryonal Sarcoma

- Affects older children & adolescents
  - 50%: 6-10yo
  - 90%: < 15yo
- Presenting signs & symptoms
  - Fever
  - Painful mass
  - Normal AFP
- Large, 7-20cm with cystic spaces
- Metastasizes to lung & bone
- Histologically
  - Primitive spindle cells resembling embryonal cells
- US
  - Variable, due to differing amounts of cystic involvement
- CT
  - Hypoattenuation with multiple septations

Embryonal Sarcoma



Primary Endodermal Sinus Tumor

- Rare, generally arise in testis/ovary
  - 10-15% extragonadal
  - Few reported cases of primary liver origin
- Germ cell origin
- >10cm
- Friable w/ necrotic/hemorrhagic areas
- Presenting signs & symptoms
  - Abdominal mass
  - Increased AFP
- CT
  - Central necrosis
  - Similar appearance to hepatoblastoma & HCC, need biopsy for diagnosis

Imaging Role in Malignant Neoplasms

- Determine extent
- Preoperative planning
  - Cure: commonly resection/transplant
- 50-60% of hepatoblstomas are resectable
- 1/3 of pediatric HCC is resectable
- Chemotherapy
  - Post-resection: adjuvant txt
  - Pre-resection: if decrease in mass, tumor resection may be subsequently feasible

Pediatric Hepatic Masses: Generalities

- Age
  - ≤ 6 months old
    - Hemangioendothelioma
  - ≤ 3 years old
    - Hepatoblastoma
    - Mesenchymal Hamartoma
    - Metastatic Disease
      - Neuroblastoma
      - Wilms' tumor
  - Older children & adolescents
    - HCC (5-15yo, 12yo median)
    - Focal Nodular Hyperplasia
    - Hepatic Adenoma
    - Embryonal sarcoma (6-10yo)
- Clinical Characteristics
  - Increased AFP
    - Hepatoblastoma
    - HCC
    - Primary endodermal sinus tumor
    - Embryonal sarcoma
  - CHF
    - hemangioendothelioma
  - Associated syndromes/disease
    - HCC
    - Hepatic Adenomas

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