Abdominal Masses of Childhood

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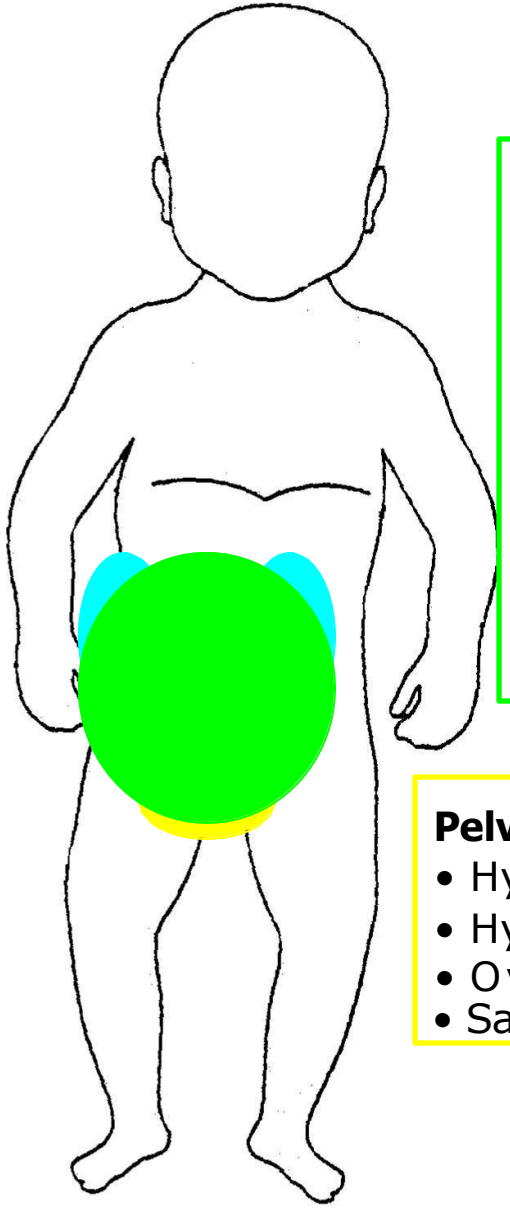
Introduction

- The finding of a palpable mass in the abdomen of a child requires immediate evaluation.
- Many of these masses are found by parents or on routine physical exam. Can be caused by organomegaly, congenital anomalies, tumors and infection.
- The patient's age, sex, and location of the mass influences the differential diagnosis. Detailed H&P are of most important.
- Determining the organ or tissue of origin and whether the mass is cystic or solid can significantly narrow the diagnostic possibilities.

- Most abdominal masses are found in children less than five years of age.
- Congenital anomalies are the leading cause of abdominal masses in neonates.
- Approximately 2/3 of **neonatal** abdominal masses are renal in origin and most commonly related to hydronephrosis or a multicystic kidney.
- Gastrointestinal congenital anomalies can also present as abdominal masses.
- Malignant tumors occur more frequently in older children.
- Neuroblastoma is the most common solid tumor in infants followed by Wilms, rhabdomyosarcoma, germ cell tumors and hepatoblastoma.

INFANTS

Flank - 65%
- **Renal - 55%**
- Hydronephrosis
- Polycystic kidney
- Mesoblastic nephroma
- Renal ectopic
- Renal vein thrombosis
- Nephroblastomatosis
- Wilms tumor
- **Nonrenal - 10%**
- Adrenal hemorrhage (Waterhouse Friderichsen syndrome)
- Neuroblastoma
- Teratoma



Intraperitoneal - 20%
- **GI Masses - 15%**
- Duplication
- Meconium ileus
- Mesenteric-omental cyst
- **Hepatobiliary - 5%**
- Hemangioendotheloma
- Hepatoblastoma
- Hepatic cyst
- Choledochal cyst
- Hydrops of gallbladder

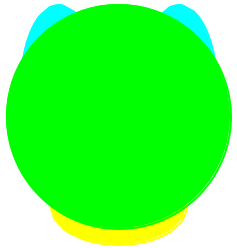
Pelvic - 15%
- Hydrometrocolpos
- Hydrocolpos
- Ovarian cyst
- Sacrococcygeal teratoma

STEPS
STANDARDIZED TOOLBOX OF EDUCATION FOR PEDIATRIC SURGERY

AMERICAN PEDIATRIC SURGICAL ASSOCIATION • FOUNDED 1970 • PATIENT CARE • RESEARCH • TEACHING

CHILDREN AND ADOLESCENTS

Flank - 78%
- **Renal – 55%**
- Wilms tumor
- Hydronephrosis
- Cystic disease
- **Nonrenal – 23%**
- Neuroblastoma
- Teratoma
- Other neoplasms



Intraperitoneal – 18%
- **GI Masses - 12%**
- Appendiceal abscess
- Intussusception
- Other neoplasms
- **Hepatobiliary – 6%**
- Hepatoblastoma
- Hepatocellular ca
- Choledochal cyst

Pelvic – 4%
- Ovarian cyst
- Hydrometrocolpos

STEPS
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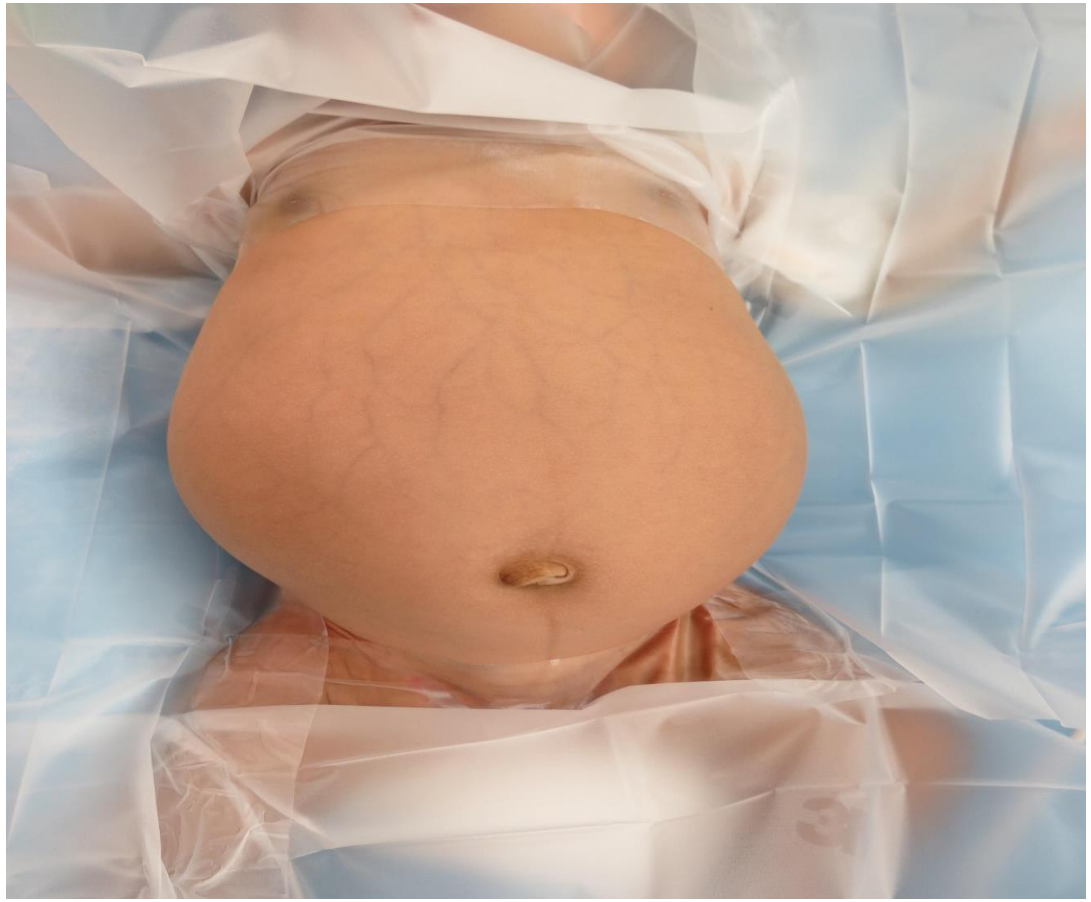
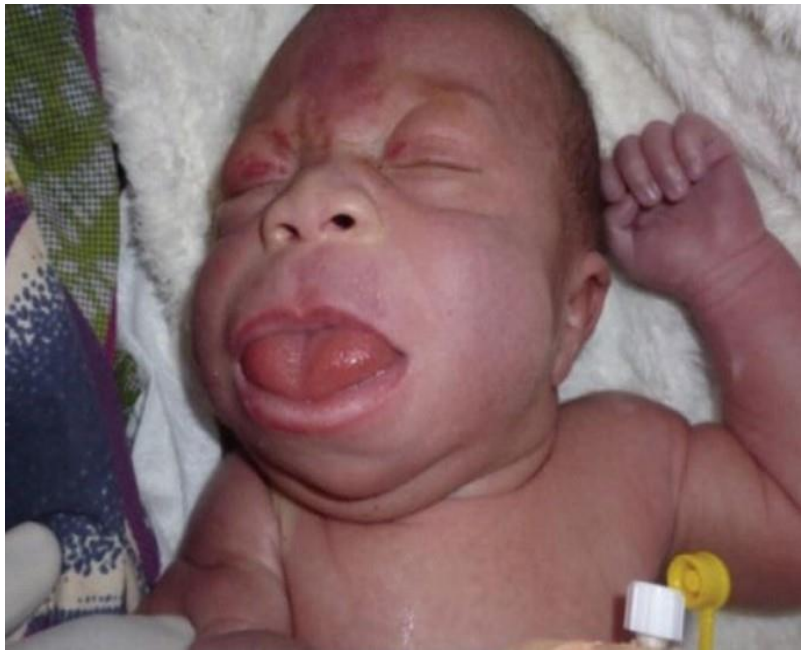


- Abdominal distension in a child with a choledochal cyst (Cyst Over the Biliary Tree)

- **Malignant masses can be associated with:**

- Fatigue, weakness, night sweat.
- Weight loss, decreased appetite (anorexia).
- Increased abdominal girth, rapid increase in size
- Decreased urination from genitourinary obstruction, constipation from gastrointestinal obstruction
- Bruising and pain on palpation or movement
- Ill appearing child with pallor, failure to thrive, cachexia and malnutrition.

BWS (Beckwith-Wiedemann syndrome)



- Macroglossia, hemihyperplasia, omphalocele, neonatal hypoglycemia, macrosomia, **embryonal tumors (e.g., Wilms tumor, hepatoblastoma, neuroblastoma, and rhabdomyosarcoma),** visceromegaly

Studies (Labs)

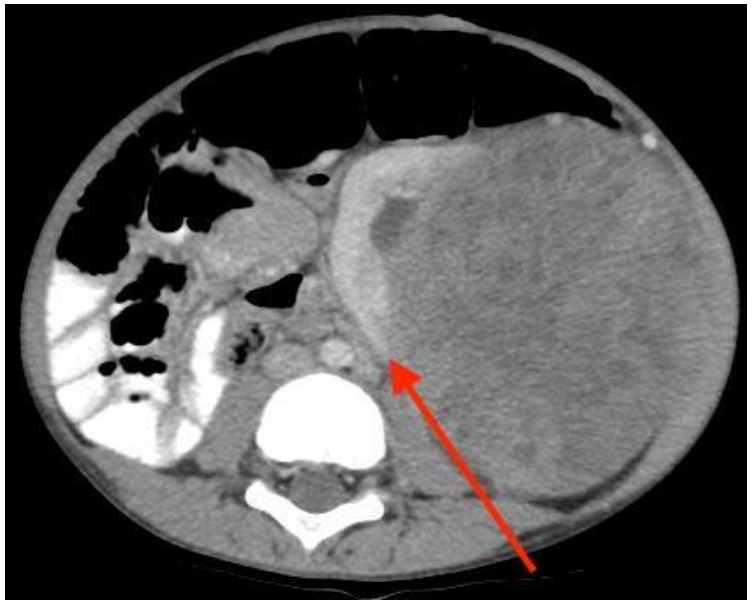
- **What labs are needed?**
  - CBC and differential
  - Lytes, BUN, Cr
  - Liver function tests
  - Amylase, lipase
  - Urine: U/A, Vanillylmandelic acid (VMA), Homovanillic acid (HVA) in NB
  - Tumor Markers: alpha-fetoprotein (AFP), β-HCG
  - LDH and Ferritin in NB

Studies (Imaging)

- **Investigations:**
  - X-rays: not usually helpful
  - US: good first test (cystic vs Soild)
  - CT: good to help plan surgery and for staging
  - MRI (SCT and HB)
  - Nuclear scans: selective use based on diagnosis. (MIBG scan in neuroblastoma)

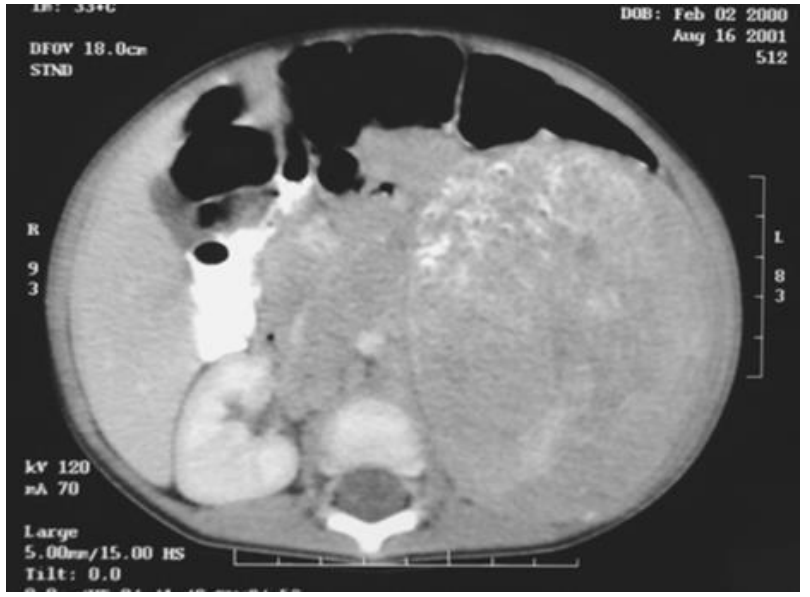
CT Scans

Wilms Tumor



- "Claw sign: Sharp angles on either sides of the mass": Wilms mass arising from kidney

Neuroblastoma



- Calcifications more likely seen in neuroblastoma

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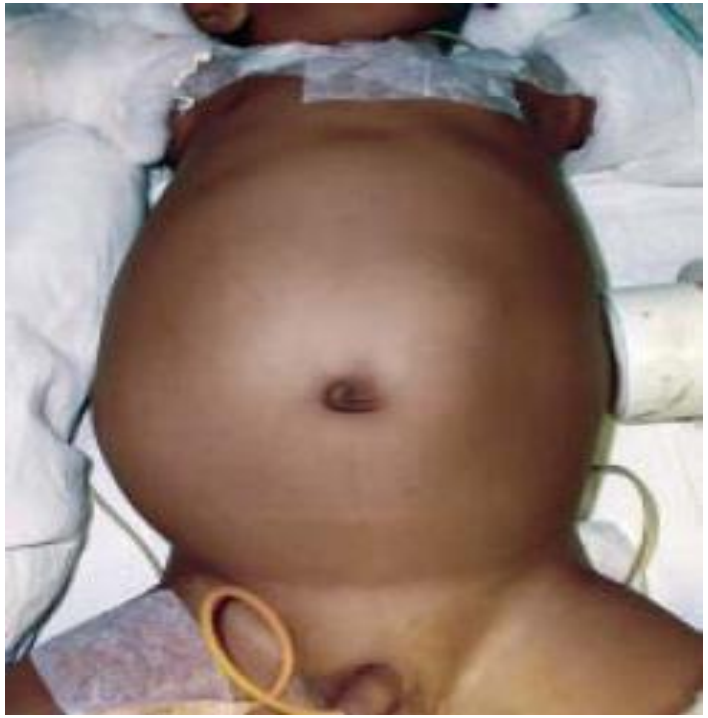
Modalities of Management

- Surgery
- Chemotherapy (neoadjuvant or adjuvant)
- Radiotherapy
- Immunotherapy

Wilms Tumor (Nephroblastoma)

- Wilms' tumor is the most common pediatric renal malignancy and 2nd most common malignant pediatric abdominal tumor after Neuroblastoma
- The mean age at diagnosis is 36 months.
- Most common presentation: asymptomatic abdominal mass, usually not tender.
- May also have hypertension, hematuria, weight loss, or anemia.
- Aniridia and hemihypertrophy is also are associated with WT
- Patients with Wilms tumors are typically healthy-appearing compared to Neuroblastoma
- May present with left sided varicocele – tumor has extended into left renal vein obstructing left testicular vein.



Staging

COG Wilms' Tumor Staging	
Stage	***Criteria***
I	The tumor is limited to the kidney and has been completely resected. The tumor was not ruptured or biopsied prior to removal. There is no penetration of the renal capsule or involvement of renal sinus vessels.
II	The tumor extends beyond the capsule of the kidney but was completely resected with no evidence of tumor at or beyond the margins of resection. There is penetration of the renal capsule or invasion of the renal sinus vessels.
III	Gross or microscopic residual tumor remains postoperatively, including inoperable tumor, positive surgical margins, tumor spillage surfaces, regional lymph node metastases, positive peritoneal cytology, or transected tumor thrombus. The tumor was ruptured or biopsied prior to removal.
IV	Hematogenous metastases or lymph node metastases outside the abdomen (e.g., lung, liver, bone, brain).
V	Bilateral renal involvement is present at diagnosis, and each side may be considered to have a stage.

- Treatment: Radical nephrectomy followed by chemotherapy +/- radiation.

- Histology and stage of tumor main prognostic indicators for Wilms tumors.

- Avoid upstaging tumor by iatrogenic complications:
  - Capsular tears, gross tumor spillage, biopsy of tumor

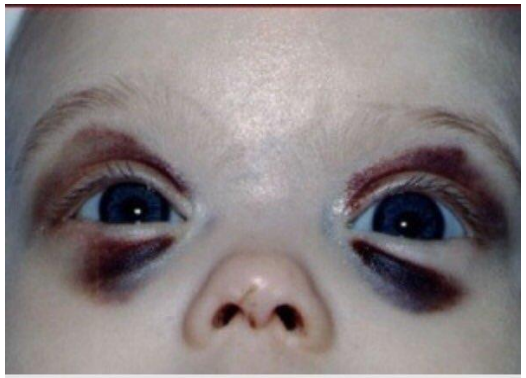
Neuroblastoma

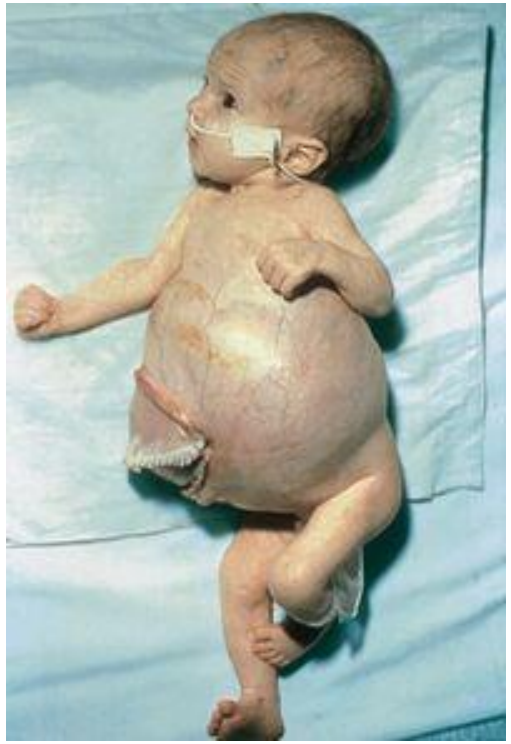
- Neuroblastoma (NB) is the *most common extracranial solid tumor* of childhood.
- Neuroblastoma is an embryonal tumor of the sympathetic nervous cells derived from the neural crest.
- Therefore, NB can originate anywhere along the path that neural crest cells migrate, including the adrenal medulla, paraspinal sympathetic ganglia in neck, chest, abdomen, and pelvis.
- 75% occurs in abdomen.
- Median age at diagnosis of 2.2 years

Presentation

- **According to primary site:**
- **Abdomen:** Painful abdominal mass
- **Neck:** Horner syndrome (ptosis, miosis, and anhydrosis)
- **Chest:** Dyspnea, Cough, Chest infection, dysphagia
- **Pelvis:** Altered defecation or urination
- Spinal cord compression signs
- Fever, wight loss, anemia, typically looks ill unlike Wilms patients.
- Racoon Eyes (retro-orbital metastasis)
- Signs and symptoms that reflect excessive catecholamine secretion include diarrhea, weight loss, hypertension, and tachycardia.

- **NB has worse prognosis comparing to WT**





Work up

- **Laboratory:**
- CBC, BMP, CMP, PT/INR, PTT, Urine VMA and HVA
- LDH, Ferritin
- AFP

- **Radiology:**
- Ultrasound
- CT scan (Chest/Abdomen/Pelvis)
- MRI
- Nuclear scan (MIBG) scan: good for bone metastasis

Table 1. International Neuroblastoma Staging System (INSS) (8)

Stage	Description
I	Localized tumor with complete gross excision, with or without microscopic residual disease; representative ipsilateral lymph nodes negative for tumor microscopically
IIA	Localized tumor with incomplete gross excision; representative ipsilateral nonadherent lymph nodes negative for tumor microscopically
IIB	Localized tumor with or without complete gross excision with ipsilateral nonadherent lymph nodes positive for tumor; enlarged contralateral lymph nodes must be negative microscopically
III	Unresectable unilateral tumor infiltrating across the midline, with or without regional lymph node involvement; localized unilateral tumor with contralateral regional lymph node involvement; midline tumor with bilateral extension by infiltration (unresectable) or by lymph node involvement
IV	Dissemination of tumor to distant lymph nodes, bone, bone marrow, liver, skin, and/or other organs (except as defined for stage 4S)
IVS	localized primary tumor (as defined for stage 1, 2A, or 2B), with dissemination limited to skin, liver, and/or bone marrow (<10% tumor) in infant younger than 1 year of age

Modalities of Management

- Surgery
- Chemotherapy (neoadjuvant or adjuvant)
- Radiotherapy
- Immunotherapy