تبييض محاضرة

Abdominal Masses of Childhood

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Done by :

لجنة الطب والجراحة

Abdominal Masses of Childhood

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STEPS
STANDARDIZED TOOLBOX OF EDUCATION FOR PEDIATRIC SURGERY

case :

(history of trauma + LUQ pain) → Pseudo-cyst at pancreas

case :

A 2-years-old M presents with abdominal mass ،What is DDX ??

Tumors محددة عند الاطفال (Wilms , neuroblastoma)

case :

A 2-days-old M presents with abdominal mass → renal masses

* Age is important in DDX

History

- **A child is seen by the PCP.** Primary care physician
- **The mother has noticed the child's abdomen was different upon bathing**

History Discussion

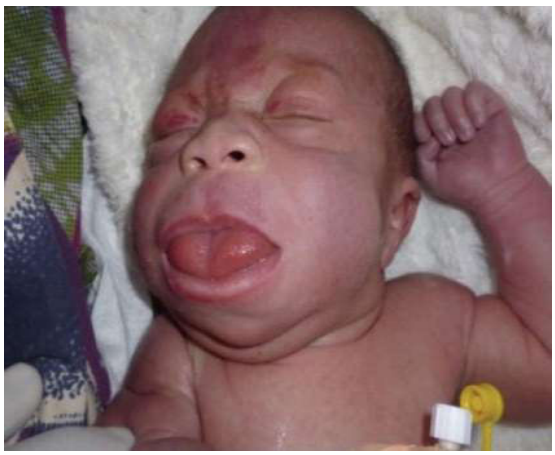
- **What other points of the history do you want to know?** Age is important for DDX so if 2 years old patient with abdominal mass think of tumours = wilms (painless) & neuroblastoma (painful) . If 2 days patient with abdominal mass think of renal masses .
  - **Age**: a crucial factor that may adjust the differential diagnosis
  - **Mass**: duration, associated pain, changes in eating and elimination patterns, history of trauma (Anorexia, wt. Loss)
  - **Birth hx**: prematurity, difficult birth, prenatal care
  - **Medical hx**: associated medical illnesses. Previous concerns for hematuria or hypertension?
  - **Family hx**: syndromes (Beckwith-Wiedemann, WAGR, Gardner)
  - **ROS**: night sweats, malaise, bleeding or bruising, skin changes

* The most common tumor in the pelvis is ((teratoma))
Sacro-coccygeal teratoma

* Common presentation = asymmetrical abdomen

Beckwith-Wiedemann syndrome

BWS



→ triad :

- large birth weight & length (macrosomia)
- overgrowth of one side of the body (hemihyperplasia)
- enlarged tongue (macroglossia)

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

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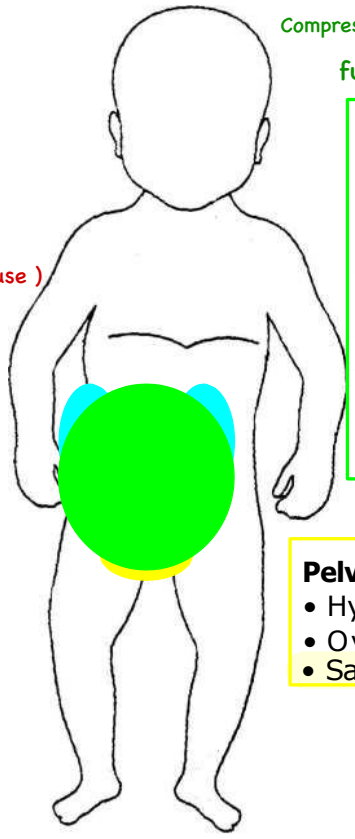
- mostly Painless: wilms tumor & Painful: neuroblastoma are the most common tumors in neonates.

INFANTS

The most common abdominal mass in infant in in flank

Flank - 65% ((Most common origin
- **Renal - 55%** of flank is renal))
- Hydronephrosis (Most common cause)
- Polycystic kidney Of renal mass
- Mesoblastic nephroma
- Renal ectopic
- Renal vein thrombosis
- Nephroblastomatosis
- Wilms tumor (Nephroblastoma)
- **Nonrenal - 10%**
- Adrenal hemorrhage (Waterhouse Friderichsen syndrome)
- Neuroblastoma Seen above the kidney



Compress the functional causes cyst or dilation

functional & non-functional part

Intraperitoneal - 20%
- **GI Masses - 15%**
- Duplication
- Meconium ileus
- Mesenteric-omental cyst
- **Hepatobiliary - 5%**
- Hemangioendotheloma
- Hepatoblastoma
- Hepatic cyst
- Choledochal cyst → cystic dilation in the biliary tree
- Hydrops of gallbladder

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

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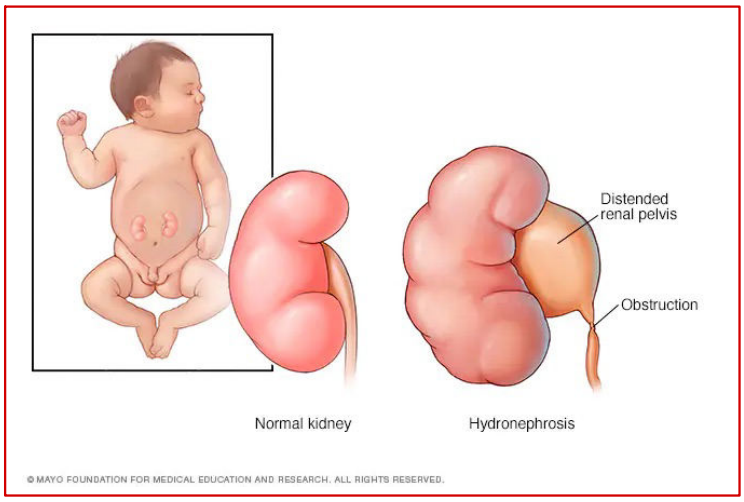
* Hydranephrosis is diagnosed by US → cystic dilation

* if there is a solid mass on US → thinK of wilms tumor

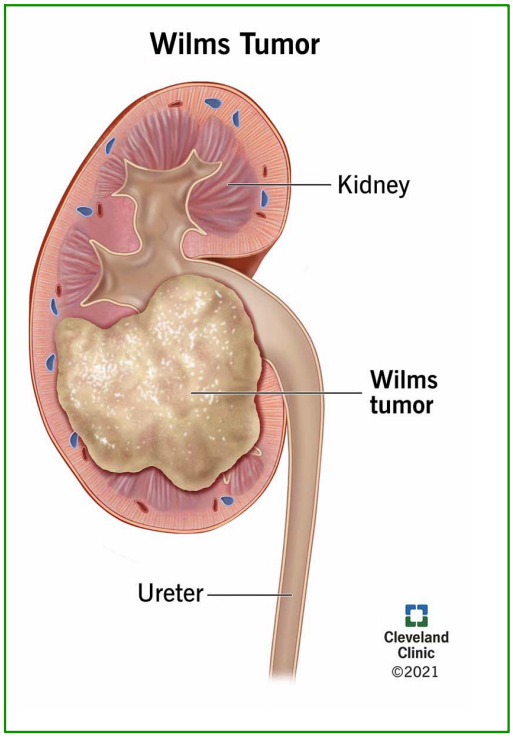


Obstruction is at the uteroplevic junction

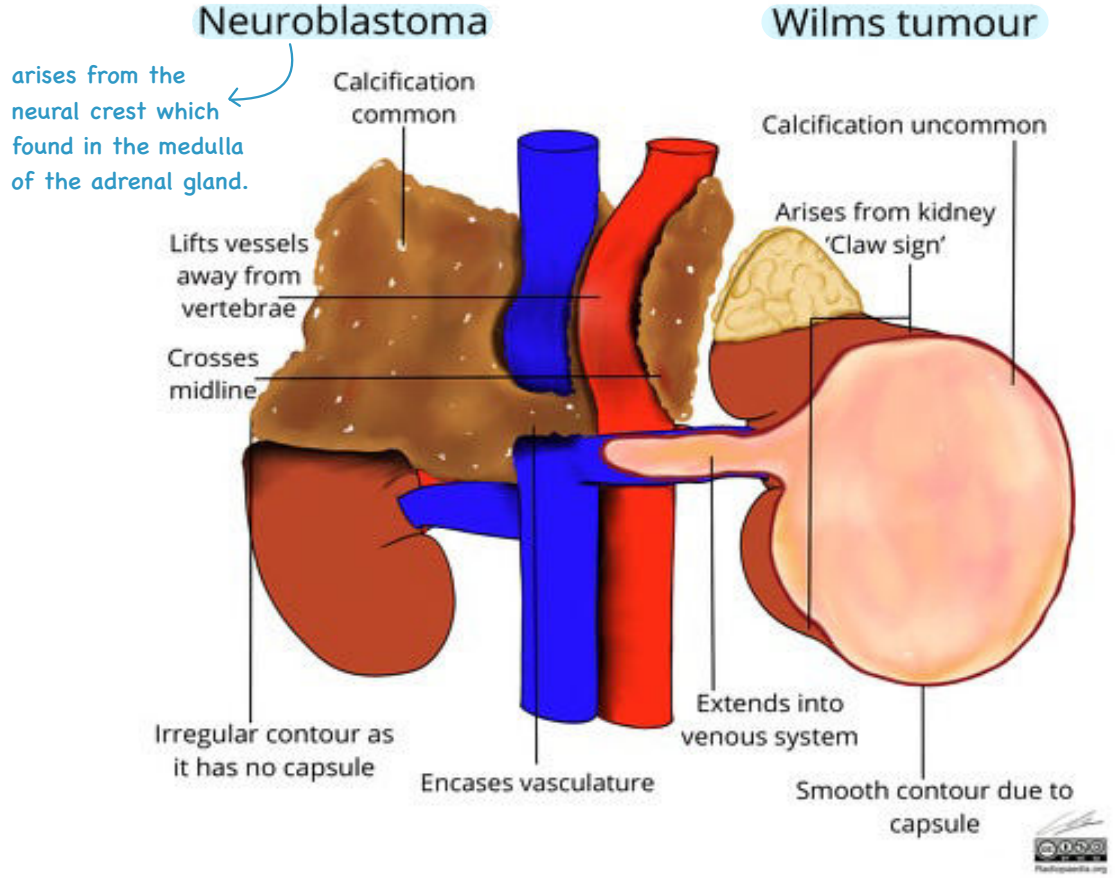
+

dilatation of the pelvis >1 cm

, Build of urine inside kidney



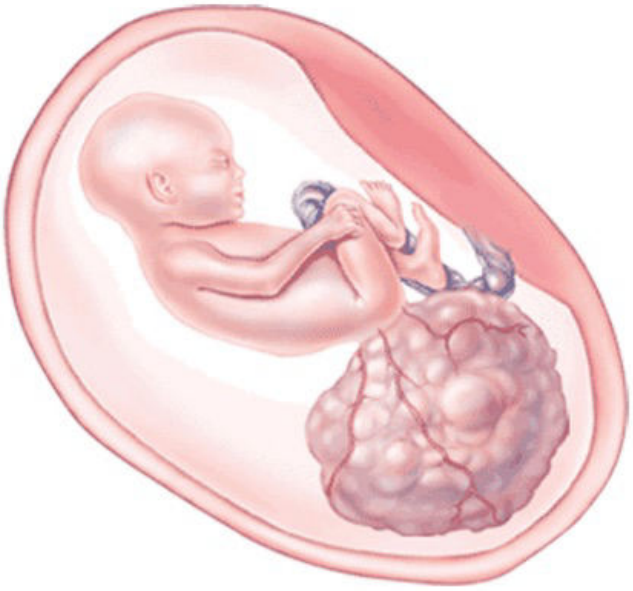
Neuroblastoma vs Wilms tumour



Sacrococcygeal teratoma (SCT) :

May compress the bowel & causes Constipation

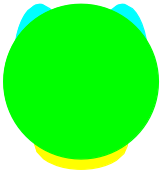
mass has tissue from endo, meso and ectoderm Layers.



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
- Other neoplasms
- **Hepatobiliary – 6%**
- Hepatoblastoma
- Hepatocellular ca
- Choledochal cyst



Pelvic – 4%
- Ovarian cyst
- Hydrometrocolpos

presence of fluid & bload in the pelvis

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Physical Exam

Such as Neuroblastoma which causes increase in secretion of catecholamine → ⬆ HR → ⬆ BP

- **What specifically would you look for?**
  - Vital Signs: some tumors can cause elevated HR, BP; some masses may push up on diaphragm and limit breathing
  - NB: This is a neoplasm of neural crest origin, arising in the adrenal medulla and along the sympathetic ganglion chain from the neck to the pelvis
  - Appearance: look for overgrowth
  - H/N: aniridia, raccoon eyes, proptosis, Horner's syndrome
  - Chest: Rapid and shallow breathing

Signs of Wilms tumor
Absence of Iris

Head & neck

Bruising around the eyes (Sign of neuroblastoma)

→ protrusion of the eye ball

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Physical Exam

- **What specifically would you look for?**
  - Cardiac: congestive heart failure (SCT=Vasular Steal Syndrome)
    - → high blood supply to the tumor
    - → usually have anemia.
  - Lymphadenopathy
  - Abdomen:
    - Omphalecele, hepatosplenomegaly
    - Mass – location, configuration, size, consistency, mobility, tenderness
  - GU: ambiguous genitalia, hypospadias, cryptorchidism ثنائية الجنس

opening of the urethra in males is not at the tip of penis.

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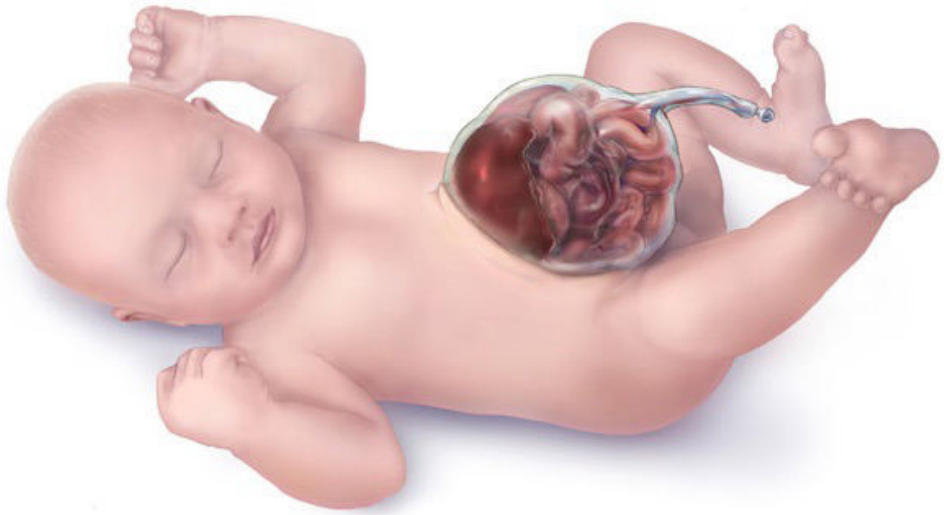
Omphalocele :

Birth defect of the abdominal wall

→ Sign for wills tumor



- abdominal mass + CHF → SCT
- abdominal mass + aniridia → wilms tumor

- abdominal mass + Raccoon eyes
- abdominal mass + Horner syndrome
- abdominal mass + proptosis

} Neuroblastoma

- abdominal mass + omphalocele → wilms tumor

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)
  - Markers: alpha-fetoprotein (AFP), β-HCG

To rule Out anemia

electro-

- Electrolytes disturbance maybe caused by cystic duplication

→ for pseudocyst

* Catecholamine

If high in the urine this indicate neuroblastoma

If high → hepatoblastoma

If high = SCT

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Studies (Imaging)

- **Investigations:**
  - X-rays: not usually helpful
  - US: good first test (the most diagnostic modality)
  - CT: good to help plan surgery and for staging
  - MRI: limited application
  - Nuclear scans: selective use based on diagnosis

To detect the origin

used in cases hydronephrosis to detect the obstruction

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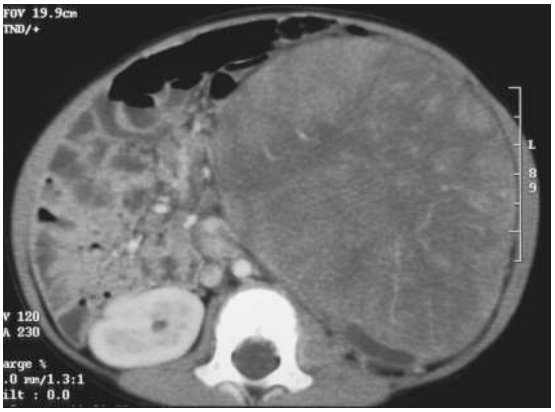
CT Scans

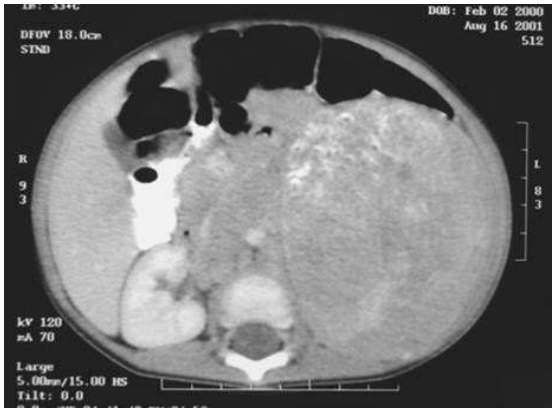
Wilms Tumor



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

Non calcified mass

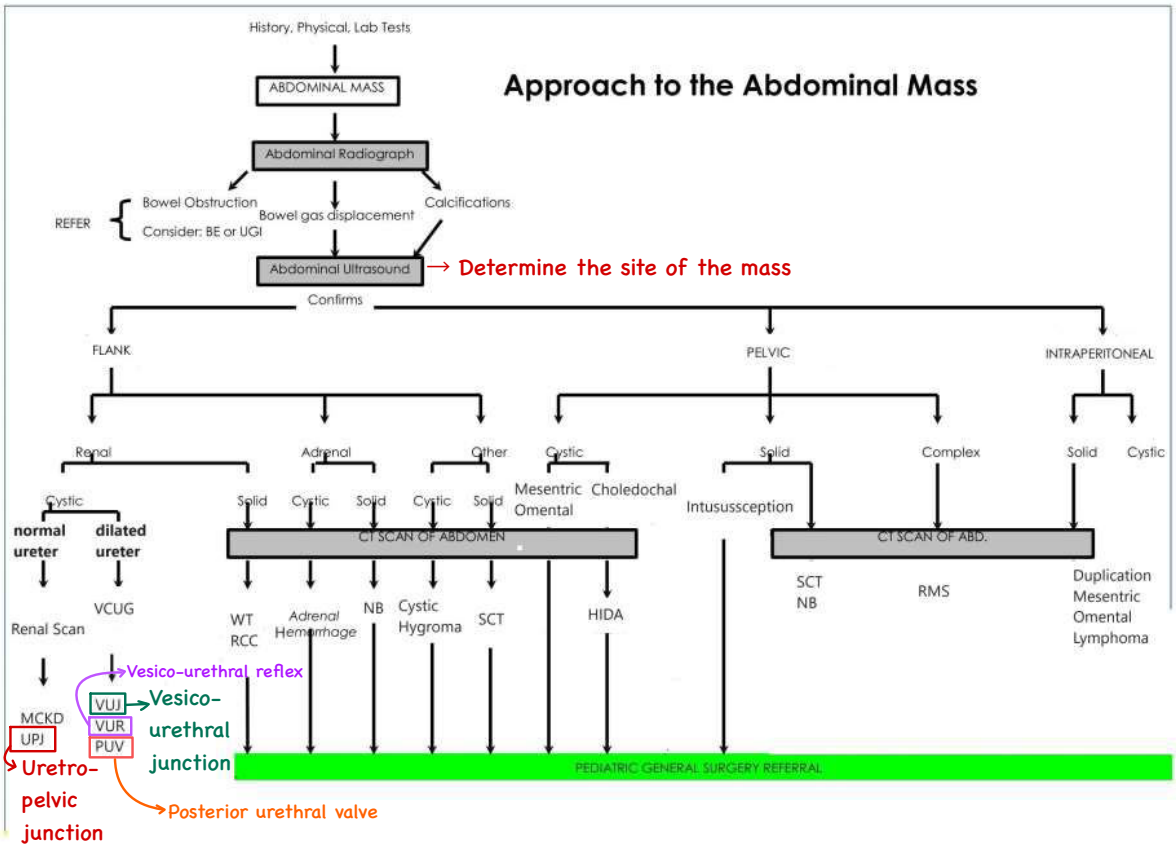
Neuroblastoma



- Calcifications more likely seen in neuroblastoma

Case Discussion

- **Diagnosis**
  - See flowchart
- **Plans**
  - See flowchart



first step is to do u/s if it's cystic dilation (hydronephrosis) if it's solid dilation (malignancy)

the next step in cystic dilation is to see the ureter if it's dilated or not , if it's dilated think of VUJO (vesico-ureteric junction obstruction) & VUR (vesico-ureteric reflux) if it's not dilated think of UPJO (ureteropelvic junction obstruction)

the next step in solid dilation is CT scan

Operation

- Goals of surgery is:
  - Staging (Invasion and LNs)
  - Obtain tissue for diagnosis only, if not resectable
  - Complete resection (avoid disruption of the margins to avoid tumor rupture) → causes upgrading of the tumor
  - Assistance with radiotherapy, or assistance with chemotherapy (Debulking)

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Complications

- **Peri-operative**
  - Ileus is common after any abdominal surgery. → caused by adhesions
  - Post-op intussusception is well reported, particularly after retroperitoneal dissection.

- **Long Term**
  - Is dependent on the tumor type and whether rupture has occurred.
  - Potential for adhesive bowel obstruction.

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Questions

1. Where do most abdominal masses arise?
   A. Flank
   B. Intraperitoneal
   C. Pelvic
   D. None of the above

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Questions

1. Where do most abdominal masses arise?
   A. Flank
   B. Intraperitoneal
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Questions

2. Which is the most useful first test to order to help determine the type of abdominal mass?
   A. X-ray
   B. Ultrasound
   C. CT scan
   D. MRI

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Questions

3. With regards to abdominal masses, the goal of surgery may include?
   A. Staging
   B. Obtain tissue for diagnosis
   C. Resection of mass
   D. Help adjuvant therapy
   E. All the above

Questions

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   A. Staging
   B. Obtain tissue for diagnosis
   C. Resection of mass
   D. Help adjuvant therapy
   E. <u>All the above</u>

Questions

4. With regards to abdominal masses, avoiding tumor rupture is critical to?

   A. Avoiding pathologic misinterpretation
   B. Intraoperative blood loss
   C. Not upstaging the patient
   D. Decreasing likelihood of postoperative intussusception
   E. Spuriously increasing tumor markers after surgery

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Questions

4. With regards to abdominal masses, avoiding tumor rupture may be critical to?

   A. Avoiding pathologic misinterpretation
   B. Intraoperative blood loss
   C. Not upstaging the patient
   D. Decreasing likelihood of postoperative intussusception
   E. Spuriously increasing tumor markers after surgery

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Review

- History and physical are key to help determine the type of abdominal mass.
- Most masses arise from the flank.
- US is the first test to do to determine the source.
- CT is the next test to help surgical planning.

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the second most common solid tumor
In the abdomen

Originates from embryonic nephrogenic multipotent mesodermal cells.

Wilms Tumor (Nephroblastoma)

Discussion of History Elements

- Congenital abnormalities?
  - Associated with WAGR syndrome, Beckwith-Wiedemann syndrome (BWS), Denys-Drash syndrome (DDS=WT+DSD), hemihypertrophy and Perlman syndrome (low muscle tone+DD)
- Abdominal pain? Painless mostly
  - Most commonly asymptomatic or no abdominal pain
- Hematuria?
  - Gross hematuria (18%)
  - Microscopic hematuria (24%)
- Weight loss/Appear ill?
  - Patients with Wilms tumors typically healthy appearing compared to Neuroblastoma – ill appearing due to typical metastatic disease
- Hypertension?
  - 20-25% of patients

Some patients are hypertensive.

WAGR :
W: Wilms tumor
A: Aniridia
G: Genetourinary
R: mental Retardation

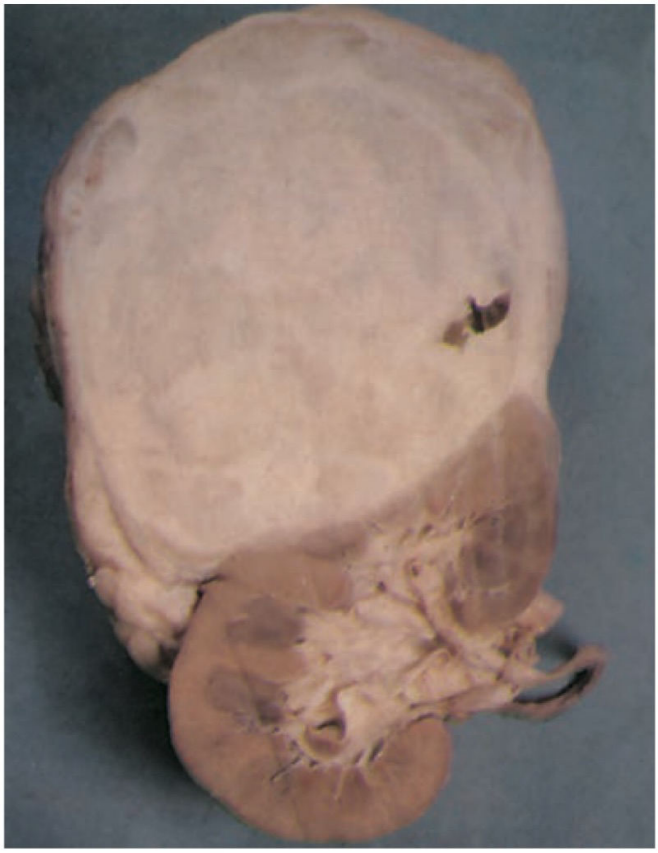

Figure 75.38 Wilms' tumour.

- The most common solid tumor in children → CNS tumors (Brain)
- The most common solid tumor in the intraperitoneal in children → neuroblastoma

& the Second most common → Wilms tumor

Physical Exam

- Healthy appearing toddler with palpable mass in abdomen/flank, non tender., asymmetrical abdomen

- Neuroblastoma → tender

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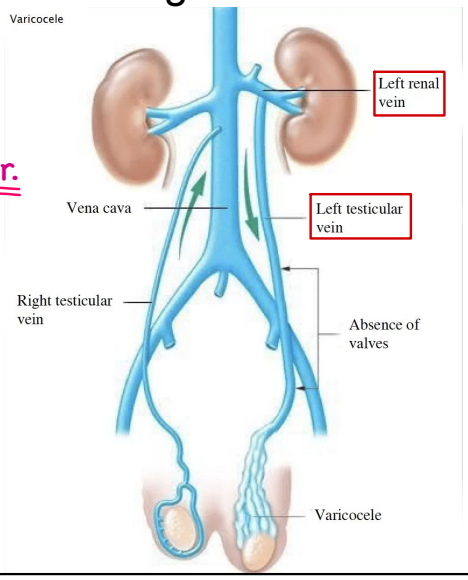
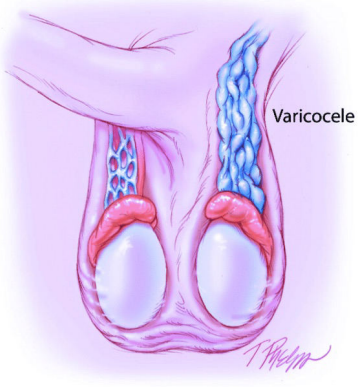
Physical Exam

- Left sided varicocele – tumor has extended into left renal vein obstructing left testicular vein → decreases venous drainage → So, causes varicocele

-any patient with varicocele, should be checked for Wilms tumor.





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Studies (Labs, Imaging)

- What labs are needed?
  - CBC, BMP (Basic Metabolic Panel), PT/INR (International normalized ratio) – rule out anemia, coagulopathy
  - Urinalysis – evaluate for hematuria
- What imaging is needed?
  - Abdominal ultrasound
    - Including doppler to evaluate mass extension into renal vein/IVC
  - CT scan abdomen/pelvis
    - Evaluate for location of disease, whether the masses are bilateral, metastatic disease.

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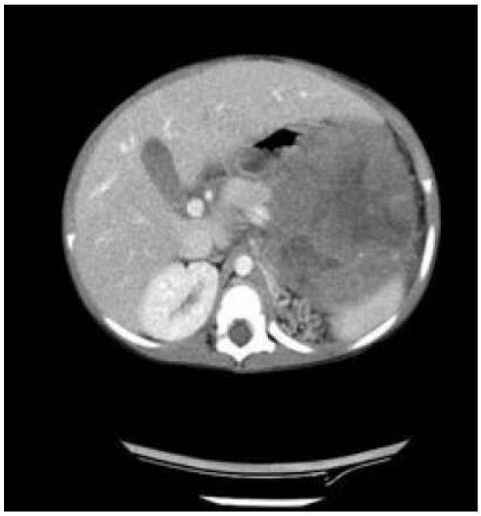
CT scan abdomen/pelvis



- Diagnosis
  - Left sided Wilms' tumor
- Management
  - Staging – two systems:
    - Children's Oncology Group (COG)
      - Focuses on surgery as primary therapy, followed by chemotherapy
    - Société Internationale d'Oncologie Pédiatrique (SIOP)
      - Focuses on neoadjuvant chemotherapy, followed by surgery

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.

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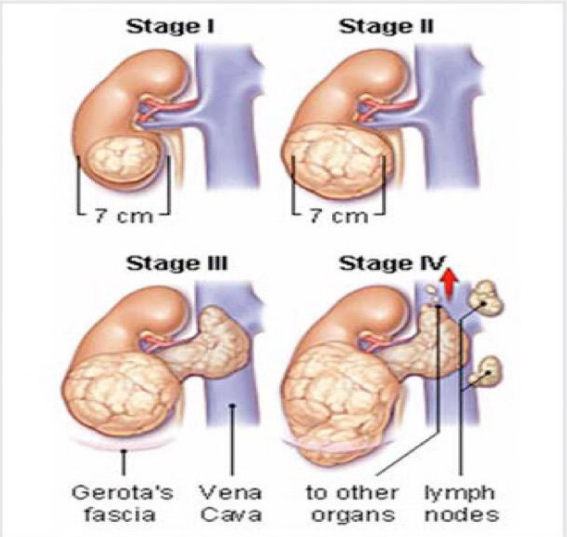
- Stage 1 (localized to kidney) has 3 years survival of >90% and cure rate is 90%.
- Stage 4 (haematogenous spread) has 3 years survival less than 30%.

Staging

•**Stage 1:**
- Limited to the kidney and completely resected



Stages of Kidney Cancer

•**Stage 2:**
- Extend beyond the kidney ,completely resected ,L.Ns do not contain tumor

•**Stage 3:**
-Tumor may be of any size and has spread to nearby major veins or L.Ns
-Deposites of tumor are found around the lining of the abdominal space
-Cancer cells are found along the edge of the removed sample
-The tumor is removed in more than one piece.

•**Stage 4:**
-the cancer has spread through the blood to organs away from the kidneys.

•**Stage 5:**
- Bilateral renal involvement at diagnosis

Case Discussion

- Management
  - Preop
    - CT scan of chest → rule out lung metastasis
      - Most common metastatic sites are lung and liver
    - Laboratory studies
      - CBC, PT/INR, PTT, Type and Screen
  - Operative
    - Radical vs partial nephrectomy

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Operation – Goals – Radical Nephrectomy

- Safely resect entire tumor
- Avoid upstaging tumor by complications
  - Capsular tears, gross tumor spillage, biopsy of tumor
- Adequately stage tumor
  - Evaluate for metastasis, removal/biopsy of appropriate lymph nodes (most common operative error), evaluating vascular invasion
- Adequately documenting pre-operative vs intraoperative tumor rupture
  - Changes post operative management with radiation

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Operation - Contraindications

- Extension of tumor thrombus above the level of the hepatic veins
- Tumor involves surrounding structures, requiring removal of those structures to remove complete tumor
- Tumor involves bilateral kidneys
- Tumor involves a solitary kidney
- Pulmonary metastasis leading to respiratory compromise

Commonly metastasis to lung 🫁

neuroblastoma عكس ال

- **All indications for neoadjuvant chemotherapy**

Shrink the tumor

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Operation – Complications

- Tumor Spill (9.7%)
  - Break in tumor capsule
    - Includes pre-operative or intraoperative needle or core biopsy (COG protocol)
    - Transection of ureter or renal vein where tumor exists
- Bleeding (2%)
- Vascular or bowel injuries

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Post-Op Management

- Peri-operative management
  - Monitor for ileus – particularly in patients with extensive retroperitoneal dissection
  - Monitor for complications:
    - Wound infection
    - Bowel obstruction
    - Intussusception
  - Prepare for radiation and chemotherapy

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Questions

The most common presenting abnormality in patient's with a Wilms' tumor is:

a. Hypertension
b. Hematuria
c. Asymptomatic abdominal mass
d. Abdominal pain

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Questions

Which of the following does NOT increase recurrence of tumor?

a. Pre-operative core needle biopsy
b. Resection lymph nodes at renal hilum, along iliac vessel and para-aortic regions
c. Transecting ureter containing tumor
d. Sustaining minor renal capsular tear during dissection from surrounding structures

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Final Discussion/Review

- Wilms' tumor is the most common pediatric renal mass and 2nd most common pediatric abdominal mass
- Most common presentation: asymptomatic abdominal mass
- Treatment for unilateral tumors: Radical nephrectomy followed by chemotherapy +/- radiation
- Histology and stage of tumor main prognostic indicators for Wilms tumors

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Final Discussion/Review

Pay attention to other associated symptoms & signs to narrow your Ddx :

1. Presence of chronic constipation = think of masses in pelvis cause it compress the bowel : sacrococcygeal teratoma SCT , somehow cause vascular steal & anaemia so do cbc & echo usually presents with congestive heart failure
2. History of trauma = develops pancreatitis & upper abdominal mass think of pseudocyst , present with high amylase levels
3. Tachycardia & hypertension = think of neuroplastoma . Elevated catecholamines , VMA , HVA
4. Vomiting & electrolytes distributed =think of duplication syndrome usually intraperitoneal
5 . Aniridia (absence of the iris) , omphalocele (abdominal wall defect) think of wilms tumor
6. Tumour markers if elevated AFP = liver tumour (hepatoplastoma) ,
elevated B-HCG = sacrococcygeal teratoma
7. On u/s if there's claw sign think of wilms tumour if there's calcification think of neuroplastoma

Wilms ::

Painless ,HTN (due to activation for angiotensin), never do biopsy (upstaging), less constitutional symptoms ,more healthy apperancer

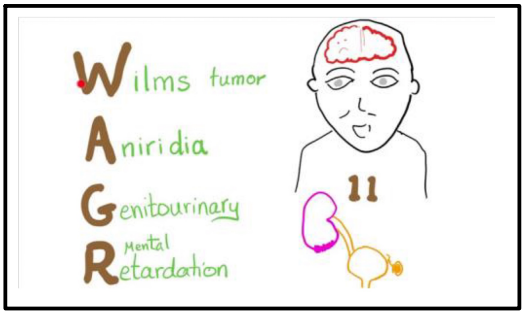
Neuroblastoma ::

painfull ,HTN (due to catecholamine release), we do biopsy then CT ,more constitutional symptoms , very sick appearance

Archive

*** Which of the following is not characteristic of WAGR syndrome? Select one:**

a. 30% risk of developing Wilm's tuour
b. Absence of the iris
c. Malformation of the genitourinary tract
d. Mental retardation
e. WT2 gene implicated



*** The main presentation of wilm's tumor is Select one:**

a. Abdominal mass.
b. Hematuria.
c. Pain.
d. Hypertension.
e. Weight loss.

*** All the statements related to Wilms' tumor are true Except:-**

a. Persistence of the varicocele when the child is supine.
b. May occur in Beckwith-Wiedemann syndrome.
c. Primary surgical resection followed by chemotherapy.
d. MRI shows the extension of the tumor to the intra-vascular.
e. Prevented cure when tumors are found in both kidneys.

*** About wilms (or abdominal masses) true except:**

Always unilateral

*** Concerning nephroblastoma (Wilm's tumor) all the following are TRUE, EXCEPT:**

A. Arising from embryonic nephrogenic tissue, so it is a mixed tumor
B. Originally it is situated in one pole of the kidney, and bilateral cases occasionally are seen
C. The most common presentation is a progressively enlarging abdominal mass noticed by the parents
D. Progressive deterioration of general health, anemia and pyrexia are common manifestations
E. The tumor spreads mainly by lymph to the para-aortic lymph nodes

* In stage 4S neuroblastoma,complete resolution of liver metastasis results from; Select one:

a. Radiotherapy

b. Chemotherapy

c. Surgery

d. Natural resolution

e. Hormonal therapy

اللهُمَّ انصُر أهل غَزَّة وثبِّت أقدامهم.

اللهُمَّ احرُس أهل غَزَّة بِعينك التي لاتَنام.

اللهُمَّ كُن لأهل غَزَّة عوناً ونصيراً، وبدِّل خوفهم أمناً وأماناً.

اللهُمَّ اجبُر كسرهم، واشفِ مَرضاهم، وتقبَّل شُهداءَهم بِرحمتكَ.

اللهم سخر لهُم ملائكة السماءِ وجُنودَ الأرض...

اللهم انصرهم على من عاداهم وافتح لهم فتحًا قريبًا.