



# **ADRENOCORTICAL CARCINOMA**

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# Abdominal Masses in Infants and Children

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- Often asymptomatic. Due to rapid tumor growth patient may have been seen by caretaker recently.
- **Benign Masses:**
  - Usually related to the GU tract, hydronephrosis, mesoblastic nephroma.
- **Malignancies:**
  - Neuroblastoma
  - Wilms tumor
  - Rhabdomyosarcoma
  - Hepatoblastoma
  - Teratoma

# Abdominal Masses in Children

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- Ultrasound, excellent screening tool.
- CT and MRI are preferred imaging modalities.
- Complete staging includes:
  - CT scan of chest, bone scan, bone marrow aspirate and biopsy.
  - Definitive diagnosis is by histologic evaluation of tissue.

# Adrenal Excess

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- **Causes of excess adrenal steroid production:**
  - Glucocorticoid excess (Cushing syndrome).
  - Mineralocorticoid excess (Conn syndrome).
  - Virilizing and feminizing adrenal tumors.
    - Most are adrenal carcinomas that produce a mixed array of androgens and glucocorticoids.
- Cushing syndrome:
  - Often is used to describe any form of glucocorticoid excess.
  - Cushing disease is hypercortisolism caused by pituitary overproduction of ACTH.
  - Among infants and children younger than 7 years, adrenal tumors predominate.

# CUSHING'S SYNDROME

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- **Signs and symptoms:**
  - **Weight gain**
  - **Poor growth**
  - **Fatigue**
  - **Delayed puberty**
  - **Bruising**
  - **Headache**
  - **Nocturia**
  - **Osteopenia**
  - **Hypertension**
  - **Plethora**
  - **Acne**
  - **Hirsutism**
  - **Striae**
  - **Buffalo hump**
  - **Delayed bone age**



# Adrenocortical Tumors

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- Rare in children and adolescents.
  - 14 new cases per year in individuals less than 20 years of age in the US.
  - Two major types:
    - Adrenal adenomas:
      - Benign neoplasms that can be functionally autonomous.
      - Hypercortisolism and hyperaldosteronism are common but seldom virilization or feminization.

# ADRENOCORTICAL TUMORS

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- **Adrenocortical carcinoma:**
  - Median age 3.2 years
  - Female predominance in most age groups
  - 90% of children have clinical evidence of an endocrine syndrome.
  - Most cases are sporadic, although several hereditary cancer syndromes include this cancer.
    - Li-Fraumeni syndrome (p53 tumor suppressor).
    - Beckwith-Wiedemann syndrome (chromosome 11).
    - MEN type 1 (chromosome 11).

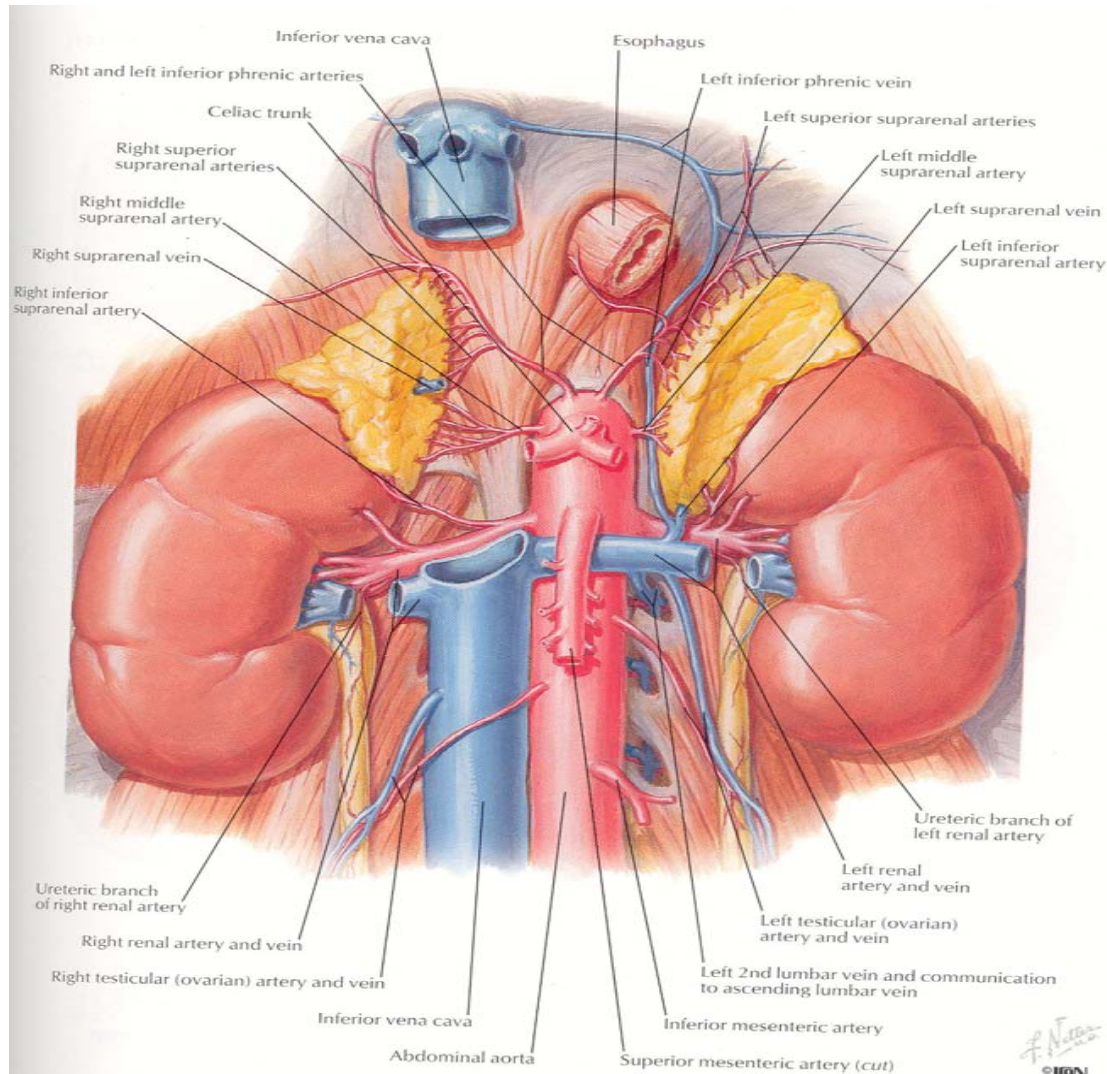
# ADRENOCORTICAL CARCINOMA

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## Clinical presentation:

- 90% of the children have evidence of an endocrine syndrome.
- **Virilization**, alone, or in combination with overproduction of other adrenal hormones is the most common presentation.

# Anatomy of Kidneys, Adrenal Glands





# OPERATIVE APPROACH

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Biopsy vs. Excision.

Open vs. laparoscopic.

# ADRENAL INSUFFICIENCY

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- Causes of adrenal insufficiency:
  - Autoimmune
  - Tuberculosis, fungal infection
  - Sepsis
  - AIDS
  - Congenital adrenal hyperplasia
  - Adrenal hemorrhage or infarction
  - Congenital adrenal hypoplasia
  - Unresponsiveness to ACTH
  - **Withdrawal from glucocorticoid therapy!**
  - Hypopituitarism
  - Hypothalamic tumors

# ADRENAL INSUFFICIENCY

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- Symptoms/Signs of adrenal insufficiency:
  - Anorexia
  - **Apathy and confusion**
  - **Dehydration**
  - Fatigue
  - **Hyperkalemia**
  - **Hypoglycemia**
  - **Hyponatremia**
  - **Hypovolemia and tachycardia**
  - Nausea and vomiting
  - Postural hypotension
  - Salt craving
  - Weakness
  - Sparse pubic and axillary hair
  - Diarrhea
  - Hyperpigmentation
  - Weight loss

