

# ADRENAL PATHOLOGY

Two distinct zones in origin and function: cortex and medulla. Arterial blood supply: variable (inferior phrenic, aorta, renal)

Venous drainage: inferior vena cava in the right side, and to the renal vein in the left

## PHYSIOLOGY:

LOCATION	ZONE	SECRETION	PATHOLOGY
Cortex (90%)	Glomerulosa	Mineralocorticoids (aldosterone)	Hiperaldosteronism Ald secreting carcinoma
	Fasciculata*	Glucocorticoids (cortisone, cortisol, corticosterone)	Adrenal Cushing Cortisol secreting carcin.
	Reticularis*	Sexual hormones (dehydroepiandrosterone, androstendione, testosterone, progesterone, estrogens)	Hiperandrogenism
Medulla (10%)		Noradrenaline 20% Adrenaline 80%	Pheochromocytoma Malignant-pheochro

### Adrenal incidentalomas:

- Non-functioning adenoma 80%
- Functioning adenoma 10%
- Malignant tumors <10%

### ALL ADRENAL INCIDENTALOMAS MUST HAVE A METABOLIC EVALUATION

Masses > 6cm high suspicion of malignancy  
Biopsy: differentiate benign- metastatic  
Functional examination of the incidentaloma: determine if it is functional or suspected of malignancy  
Clinical, hormonal and radiological evaluation.

\*Overlap can occur with hormone synthesis and pathologies between fasciculata and reticularis

### HIPERCOTISOLISM – CUSHING’S SYNDROME

**-Suspect:** Stretch marks, fragile capillary, acne, hirsutism, central obesity, hump, full moon-face, HT, muscular weakness

**-Causes:**

ACTH.dependent 80%	ACTH.independent 20%	Pseudo-Cushing
-Cushing’s disease -Ectopic secretion ACTH or CRH (lung, thymus, pancreas)	-Adrenal adenoma -Adrenal carcinoma -Micro/macronodular hyperplasia	-Major depression -Alcoholism

**-Diagnostic tests: Screening:**

- Dexamethasone 1mg suppression: + if cortisol >1.8mcg/dL(50nmol)
- Nocturnal salivary cortisol: Positive if elevated
- Free urine cortisol (24 hours): Positive if >3x normal

**Confirmation:**

- Nocturnal salivary cortisol
- Weak dexamethasone 2mg suppression test + CRH stimulus

### PRIMARY HYPERALDOSTERONISM – CONN’S SYNDROME

**-Suspect:** HYPERTENSION (HT) + HipoK+, HT + adrenal mass, HT>150/100 3 different times in 3 days, HT not controlled by three medications

**-Causes:** Bilateral idiopathic hyperplasia: 65-75%  
Aldosterone producing adenoma: 25-35%

**-Diagnostic tests**

- 1st: **Plasmatic aldosterone/renin ratio (ARR):**  
Positive if ARR ≥20 + aldosterone >15ng/ml  
K sparing diuretics must be stopped before test
- 2nd: **Confirmation tests:**  
Oral overload with Na (6g/d for 3 days -> 24h urine)  
IV overload with Na (2000cc 0,9% for 4h -> plasma)  
Fludocortisone suppression test (0,1mg c/6h for 4 days)
- 3rd: **localization: unilateral vs bilateral**  
Diagnosis if unilateral adenoma 1-2cm  
If doubts, adrenal vein catheterism

**-Treatment:** Unilateral: surgical resection  
Bilateral: spironolactone, eplerenone

### ADRENAL CARCINOMA

**-Generalities.** Rare and aggressive (0.05-0.2% of all cancers)

- Adrenal cortical carcinoma functional in 80% of times
- 2-6% are bilateral and associated to hereditary syndromes: Li-Fraumeni, Beckwith-Wiedemann, MEN-1, McCune-Albright and, Carney complex

**-Most frequent presentation: Cushing’s Sd (33-53%), Cushing’s Sd with virilization (20-24%), virilization only (10-20%), feminization (6-10%)**

**-Treatment**

- Surgical resection. Limit between choosing open or laparoscopic/robot assisted: 6cm
- If M+: Mitotane (response in 20-40%, symptom relief in 70%)
- Agents used to alleviate severe symptoms: Metyrapone, aminoglutethamide, ketoconazole

### PHEOCHROMOCYTOMA

The 10%’s rule: 10% bilateral/multiple. 10% children. 10% extraadrenal. 10% malignant. 10% recur after surgery. 10% incidentalomas

**-Generalities:** Medulla: Pheochromocytoma. Sympathetic ganglia: Paraganglioma.  
Lifetime surveillance: up to 16% recurrences  
Around 25% associated to hereditary syndromes: MEN2A, MEN2B, VHL, NF-1.

**-Presentation: TYPICAL TRIAD IN 30-50%: HEADACHE, PALPITATIONS, SWEATING.**  
Paroxistic hypertension 30-50%. HT crisis induced by anesthesia, labor, biopsies, thiamine

**-Diagnosis. 1 Screening:** Fractionated plasma metanephrines (99% sensitivity)

**-2 Confirmation:** 24-h urine metanephrines and catecholamines (99% specificity)

**-3 Localization: CT:** heterogeneous mass with necrosis/hemorrhage, >10UH without contrast, >100UH with contrast

- **MRI:** T1: low intensity while in T2: high intensity (lightbulb sign)
- **MIBG:** scintigraphy is used if suspicion of extra adrenal foci

**-Treatment. Surgical resection. Vein must be ligated first in all cases**

**Surgical pre-op preparation:** 1º Alpha block: fenoxibenzamina for 2 weeks  
2º Beta block: propranolol, 3º Hydration: IV saline

**-Minimally invasive (lab/robotic) if <6cm.**

**-Post-op:** if benign and asymptomatic, no more treatment

## RADIOLOGIC PRINCIPLES OF ADRENAL PATHOLOGY

	ADENOMA	CORTICAL CARCINOMA	PHEOCHROMOCYTOMA	MYELOLIPOMA
<b>CT without contrast</b>	If <10 UH: Adenoma If <4cm: benign	>6cm in 90% >25UH Heterogeneous, irregular With calcifications and necrosis	>10UH without contrast Heterogeneous With hemorrhage and necrosis	<0 UH Fat and hematopoietic tissue
<b>CT with contrast</b>	Fast washout (>50% 10-15min)	Washout <40% in 10-15min Can be confused with metastases or lipid-poor adrenal adenoma (20-40UH, washout >50%)	>100UH with contrast	
<b>MRI</b>	T1: Loss of signal is out of sequence phase	*Only if CT is inconclusive: T2: high intensity, heterogeneous if contrast	T1: Low intensity T2: High intensity (lightbulb sign) in fat suppression phase	T1: High intensity, loss of signal in out of phase sequence T2: High intensity