# ---SURGICAL ENDOCRINOLOGY---

## I. THYROID

Follicular cells produce and store T3, T4
Parafollicular cells (C cells) secrete calcitonin

### Thyroid nodule

4% of population
4:1 female:male ratio (but ↑risk of malig in men)
85% are benign
Malig risk factors: childhood radiation, solitary nod, cold nod, compressive sxs
Dx: TSH, Ca levels (r/o MEN), calcitonin
- FNA = most important test (only 3% false neg)
- Follic adenoma diff to disting from follicular carc on FNA
  - May req excision for dx
- Mult nodules are usu benign but may req excision for dx
- Most cystic masses resolve after FNA

U/S→cystic→FNA→drain
↓
solid (or residual cyst)→FNA→benign→T4 suppr'n
↓
Indet/malig→excise

Rx: Benign nodules: 3-6 months of T4 suppr'n therapy
- If nod shrinks, cont T4; if no change in size, reaspirate; if enlarges, excise
- Initial surgical proced I/L thyroid lobectomy + isthmectomy
  - If init path non-dx, await final path results before doing total thyroidect

Cx: Injury to parathyroid gland or blood supply
- Recurrent laryngeal nerve inj (transient neuropraxia more common, resolves)
- Hematoma

### Hyperthyroidism

Lifetime risk 5% for females, 1% for males
Grave's dz #1 cz
- Exophthalmos, pretibial myxedema (pathogenesis unknown)
- Rx: 1. PTU, methimazole (inhib thyroid hormone synth)
  - 1/3 of pts remain in remission at 1yr but 5-10% have adverse side effects
  - BB decrease periph conversion of T4 to T3 (for short term management)
  - 2. Radioiodine ablation: safe and effective
  - 3. Thyroidectomy: best for reduction of exophth and myxedema

Other czs: toxic adenoma, toxic multinod goiter, ovarian teratoma
Toxic adenoma
- No associated ophthalmopathy or myxedema
- Nodule is "hot" on thyroid scan
- Rx: Ablation has high recurrence rate
  - Lobectomy and isthmectomy is optimal

**Thyroid cancer**

2:1 female:male ratio

Surgery is optimal Rx, except for anaplastic type or lymphoma

1. Papillary carcinoma
    - #1 type (70-80% of cases)
    - Often in young females
    - Psammoma bodies, orphan annie cells
    - Good Px for most; worse px if male, >50 yo, >4cm
    - Rx: Total thyroidectomy + postop radioiodine ablation
        - All pts also need T4 to suppress TSH dependent tumors
        - Mets to lung, bones: rx w/ radioI ablation after thyroidectomy
        - May need additional external radiation for bone mets
2. Follicular carcinoma
    - 10-20% of all cases
    - Often in females in 50s
    - Resembles follicular adenoma on cytology (need bx to show capsule/vasc invasion)
    - Mets spread hematogenously to lung and bone
    - Rx: same as for papillary carcinoma
3. Medullary carcinoma
    - 7% of all cases (20% of these genetic)
    - Always B/L in Familial MTC and MEN 2
    - Calcitonin levels elevated (check urine metaneph to r/o pheo)
    - Rx: Total thyroidectomy + central LN dissection
    - Px: 50% 10 yr survival
4. Anaplastic carcinoma
    - Very aggressive
    - Elderly pts
    - 0% 2 yr survival
    - Rx: surgical resection for palliation; chemo, radiation ineffective

**II. PARATHYROID**

PTH: N terminal is active segment; t½ = 3mins

Vit D activation: skin→liver→kidney

Hypercalcemia: cz: "CHIMPANZEES"; Sxs: "stones, bones, moans, psychic overtones"

Most pts have minimal sxs, often found to have ↑Ca incidentally

Most common sxs are vague, constitutional sxs or psych changes (depression)

Dx: ↑PTH, Ca, Cl; ↓Phos, HCO3; Cl:Phos ratio >33; ↑urine camp, Ca

↑PTH occurs w/: primary hyperPTH, familial hyperCa hypocalciuria, vit D def

Primary hyperPTH
- #1 cz of outpt hyperCa
- 85% d/t parath adenoma, 15% parath hyperplasia, <1% parath carcinoma
- Homeostatic set point for Ca is reset at higher value

Secondary hyperPTH
- Usu renal failure pts (↓vit D synthesis)

Also d/t ↓vit D intake, ↓sun exposure
Tertiary hyperPTH
Develops in pts w/ secondary hyperPTH
Hyperplastic glands become autonomous producers of PTH
PTH remains ↑'d even w/ renal transplant and correction of Ca/Phos levels
Rx: Acute hyperCa: IVF, loop diuretics
ESRD pts: vit D, phosphate binders, parathyroidect occasionally, renal transplant
Surgery recommended for even minimal sxs
Single gland excision for adenoma
Subtotal parathyroidect (3 ½ glands) for secondary and tertiary hyperPTH
Preop localization w/ sestamibi radionuclide scan or U/S
Intraop PTH (ioPTH) monitoring technique
Check PTH levels q5mins after excision; should ↓>50% at 10 mins
Cx: Recurrent laryngeal N injury
Transient post op hypoPTH d/t gland suppression (↑'d risk in pts w/ bone dz)
Hungry bone synd: Ca/Phos remain low d/t take up by bone
Give large doses of Ca/Phos until synd resolves
Parathyroid carcinoma
Young pts
One palpable gland
Invasive, recurs, thus poor px
Rx: I/L lobectomy w/ LN dissection

## III. ADRENALS

Superior, middle, inferior adrenal arteries arise from inf phrenic, aorta, renal artery
R adrenal v into IVC, L into L renal v
Cortex: zona glomerulosa, fasiculata, reticularis
Medulla: NE, EPI

### Cushing's syndrome

4:1 female:male ratio
#1 cz = exogenous steroids
Cushing's dz: ACTH-producing pituitary adenoma (#1 endogenous cz)
Dx: 24 hr urine cortisol/17OHCS, overnight dex suppression test to confirm Cushing's
Then measure ACTH and check hi dose dex suppression
Low ACTH = adrenal tumor
Hi ACTH w/ no suppression = ectopic ACTH (f/u CT chest)
Hi/nl ACTH w/ suppression = pituitary ACTH (f/u MRI)
Rx: Metyrapone, ketoconazole (steroid inhibitors)
For pituitary adenoma: transphenoidal resxn (95% initial cure, 50% recurrence)
B/L adrenalectomy if pituitary rx fails; Cx: enlargement of pituitary adenoma
Primary adrenal hypercortisolism: 90% d/t solitary adenomas
Remaining adrenal tissue becomes atrophic
Periop and postop (12-18 mo) of steroids required (ACTH stimulation test before d/c)
Laparascopic partial adrenalectomy = best approach

**Conn's syndrome**
d/t solitary adenoma, nodular hyperplasia, or diffuse hyperplasia
2:1 female:male ratio
Triad of HTN, hypoK, hi aldo / low rennin
Sodium load test used to distinguish b/t primary and secondary hyperaldo
May also use venous sampling of aldo levels
If cz of hyperaldo not localized to one adrenal gland, likely d/t B/L adrenal hyperplasia
    Manage w/ spironolactone

**Adrenal cortical carcinoma**
Rare, slight left side and female predominance
Most are large (mean 12cm)
50% pts p/w Cushing's synd, 15% w/ virilization
50% have mets at dx (extension thru adrenal vein)
Dx: abd CT
Rx: resection usu poss, w/ 5 yr survival of 55%
Post op steroids usu needed

**Incidentalomas**
33% are benign adrenal cortex adenomas, 22% are mets
Increased risk of hyperfunction if >4cm
Increased risk of malignancy if >6cm
Dx: 1st determine if hormonally active; 2nd if malignant
    1mg dex suppression test, plasma-free metaneph levels, aldo:renin ratio
    #1 cancer = mets from lung, breast, colon
    FNAB; benign if clear fluid aspirate
Rx: If <4cm: may follow w/ abd CT in 6 months
    If >6cm: excise d/t malignancy risk

**Pheochromocytoma**
85% from adrenal medulla (R>L)
Rule of 10s (10% each): familial, malig, B/L, extra adrenal (paraganglionoma)
Triad of Sxs: episodic HTN w/ h/a, palpitations, sweating
Dx: plasma-free metaneph/normet; MRI for localization
Rx: 1st step: phenoxybenzamine (alpha antagonist) then excision
    Periop steroids may be necessary d/t adrenal suppression

**Multiple endocrine neoplasms (MEN) 1**
Screen 1st degree relatives of MEN pts Q3 yrs
Check Ca and PTH levels in all pts w/ islet cell tumors (hi sensitivity)
1. Parathyroid hyperplasia
    #1 d/o in MEN 1 (90% pts)
    Mean age of onset 25 yo
    Recurrence rate s/p parathyroidectomy is 50% at 10 yrs
2. Pancreatic islet cell dz

#1 islet cell neoplasm is nonfunctional, secreting panc polypeptide
#1 functional tumor is gastrinoma (often in prox panc or duod wall)
May also secrete insulin, glucagons, VIP, somatostatin
Most insulinomas are benign; other islet neoplasms are mostly malignant
50% pts have liver mets (except w/ insulinoma)
Direct correlation b/t tumor size and mets
Tend to grow more slowly than most GI cancers
Rx: excision (gastrinomas usu mult, req Whipple)
Parathyroidectomy should precede gastrinonma resection

3. Pituitary dz
Almost all secrete prolactin (irreg menses, galactorrhea, infertility)
Dx: MRI or CT of sella, measurement of prolactin levels
Rx: bromocriptine, resection

**Multiple endocrine neoplasms (MEN) 2**

Mutation in RET proto oncogene
RET testing should be done all MTC and pheochromo pts
Medullary thyroid cancer (MTC)
MEN 2A and 2B pts
Arises from parafollicular, C cells (calcitonin secreting)
Screen for pheo (and if +, perform adrenalectomy) before surgery for MTC

MEN 2A
RET + pts should have total thyroidectomy by age 6 yo to prevent MTC
50% MEN 2A pts have pheo
In U/L dz, 50% risk of developing C/L dz
25% MEN 2A pts have primary hyperPTH (less severe than MEN 1)

MEN 2B
MTC most aggressive in MEN 2B
In RET + pts, perform thyroidectomy before age 6 months
Marfanoid habitus

Familial MTC
Thyroidectomy in childhood (similar to MEN 2A)