ADRENAL GLAND SURGERY

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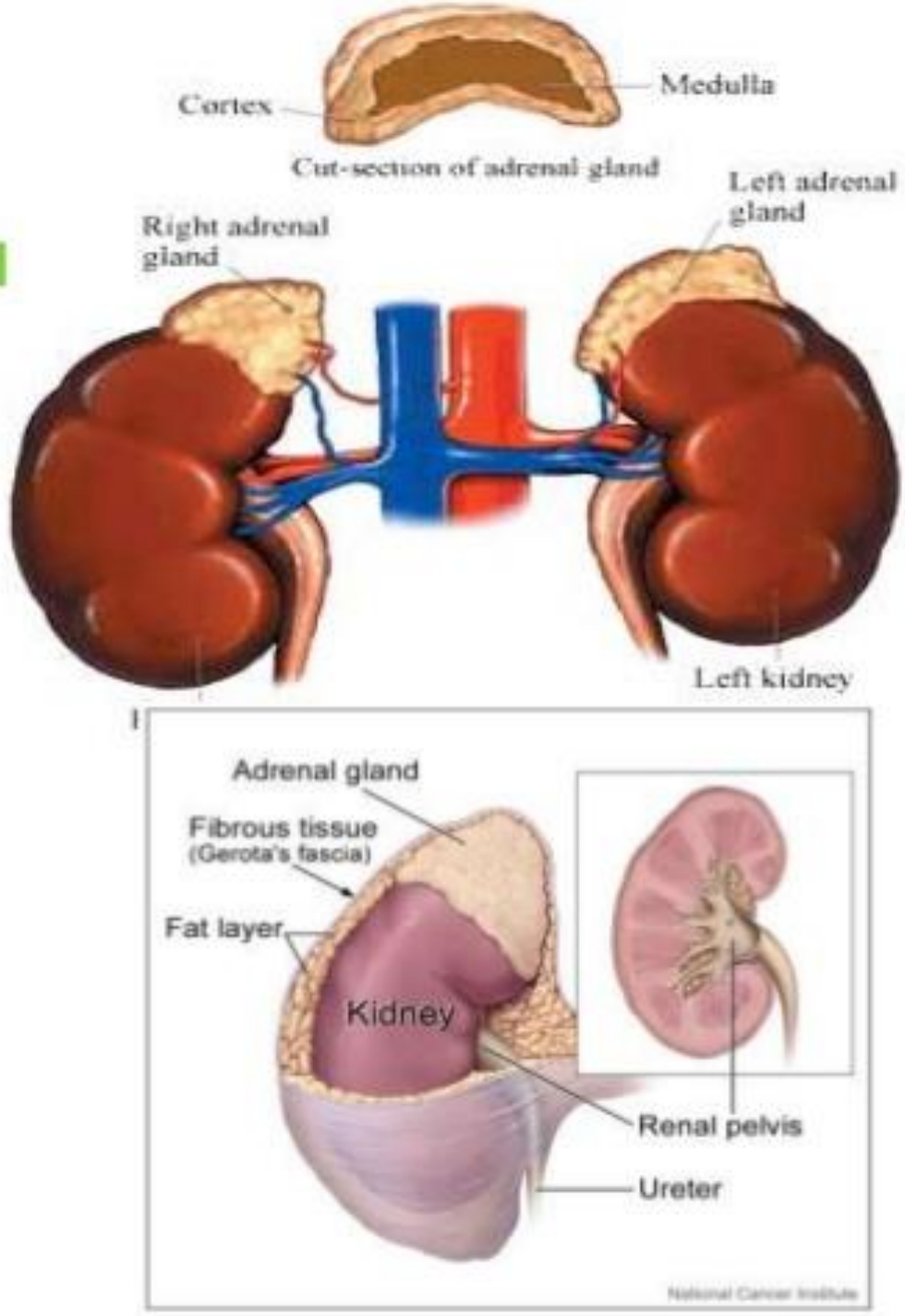
OBJECTIVES

- TO UNDERSTAND:
- The anatomy and function of adrenal and other endocrine glands
- The diagnosis and management
- The role of surgery

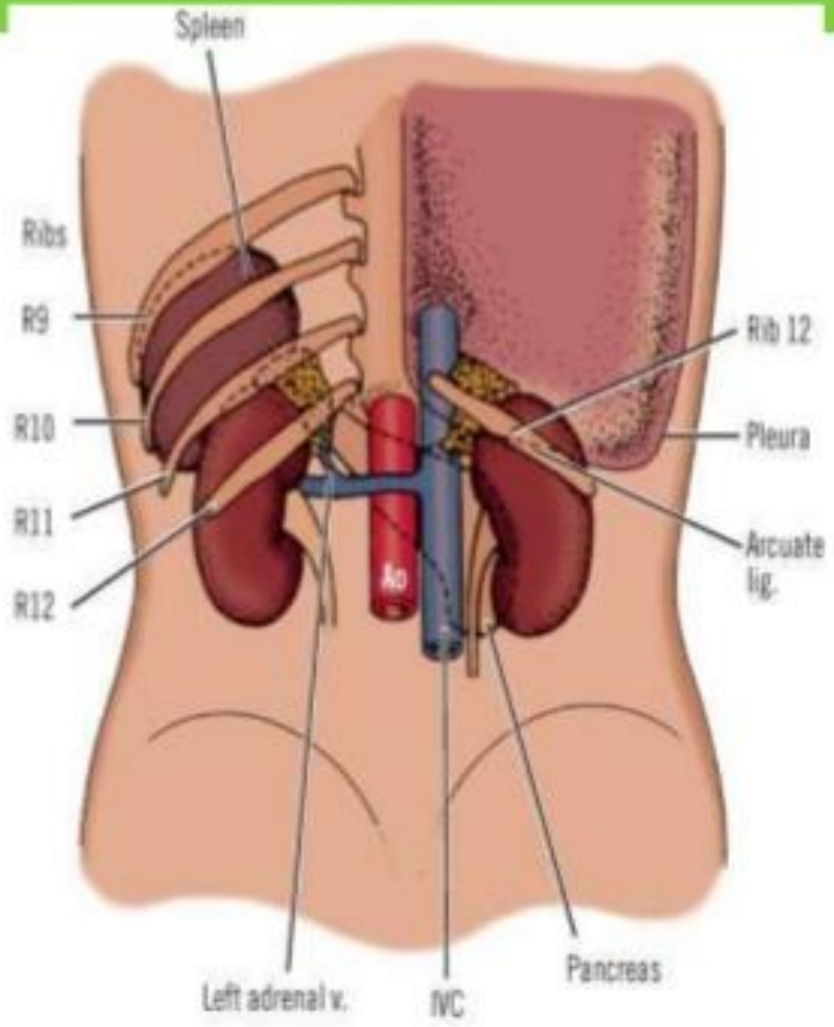
ANATOMY

- Weight = 4g
- 2 adrenal gland, right and left
- 2 component ; inner adrenal medulla and outer adrenal cortex
- Situated near upper poles of kidneys in retroperitoneum, within Gerota's capsule



ANATOMY

- Right adrenal gland – between right liver lobe and diaphragm
- Left adrenal gland – close to upper pole of left kidney and renal pedicle, covered by pancreatic tail and spleen



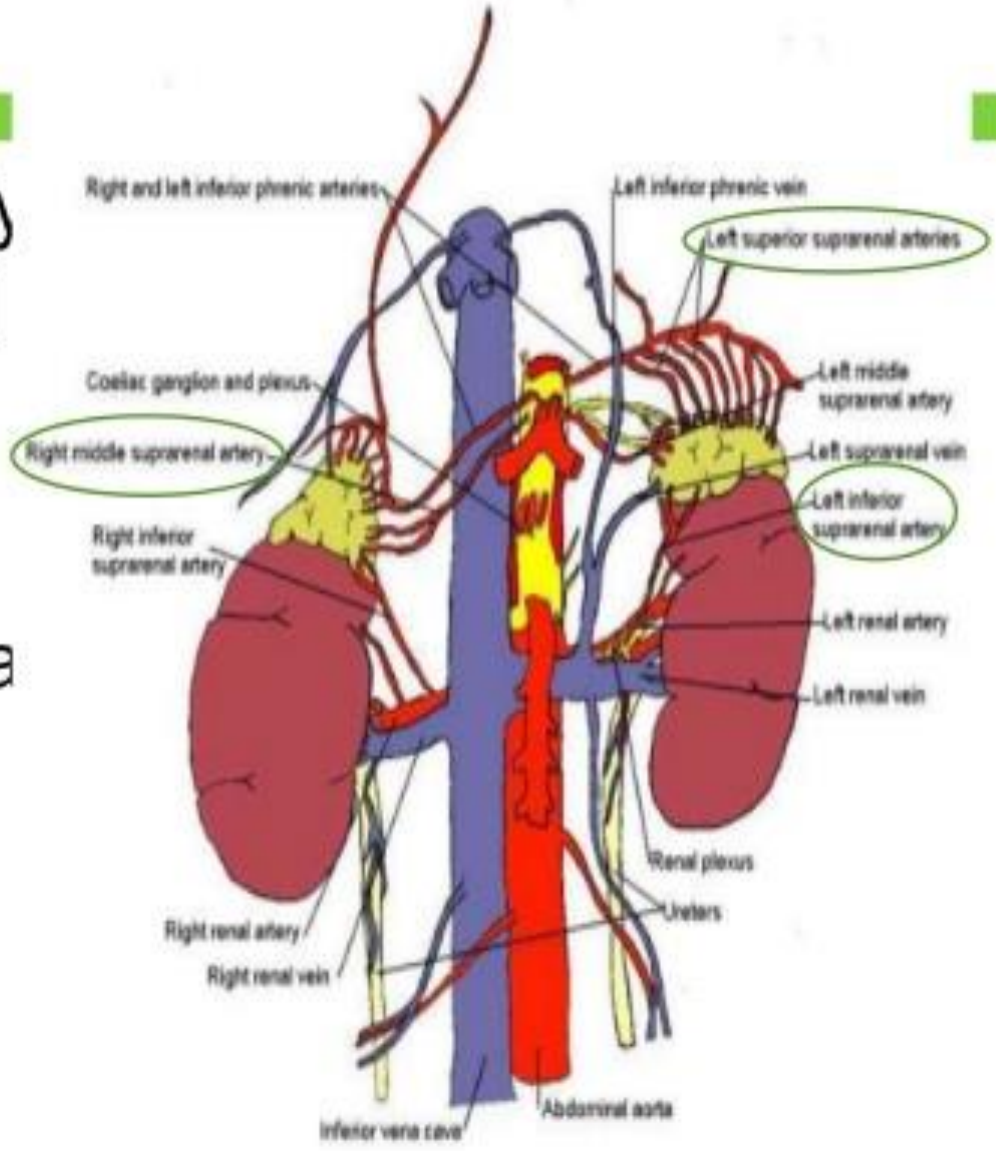
ANATOMY

- Arterial blood supply

–superior suprarenal artery (from inferior phrenic artery)

-middle suprarenal artery (from abdomina aorta)

- inferior suprarenal artery (from renal artery)

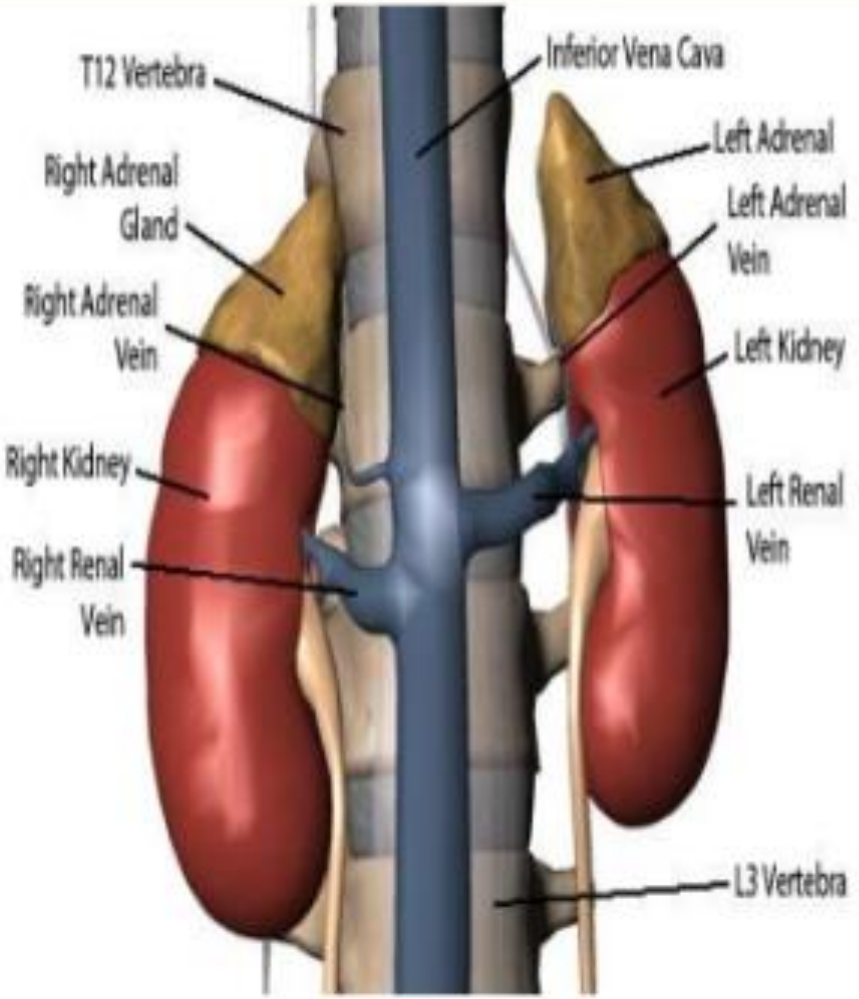


ANATOMY

- Venous blood supply

–Right suprarenal veins drain into inferior vena cava

- Left suprarenal vein drain into left renal vein or left inferior phrenic vein



EMBRYOLOGY

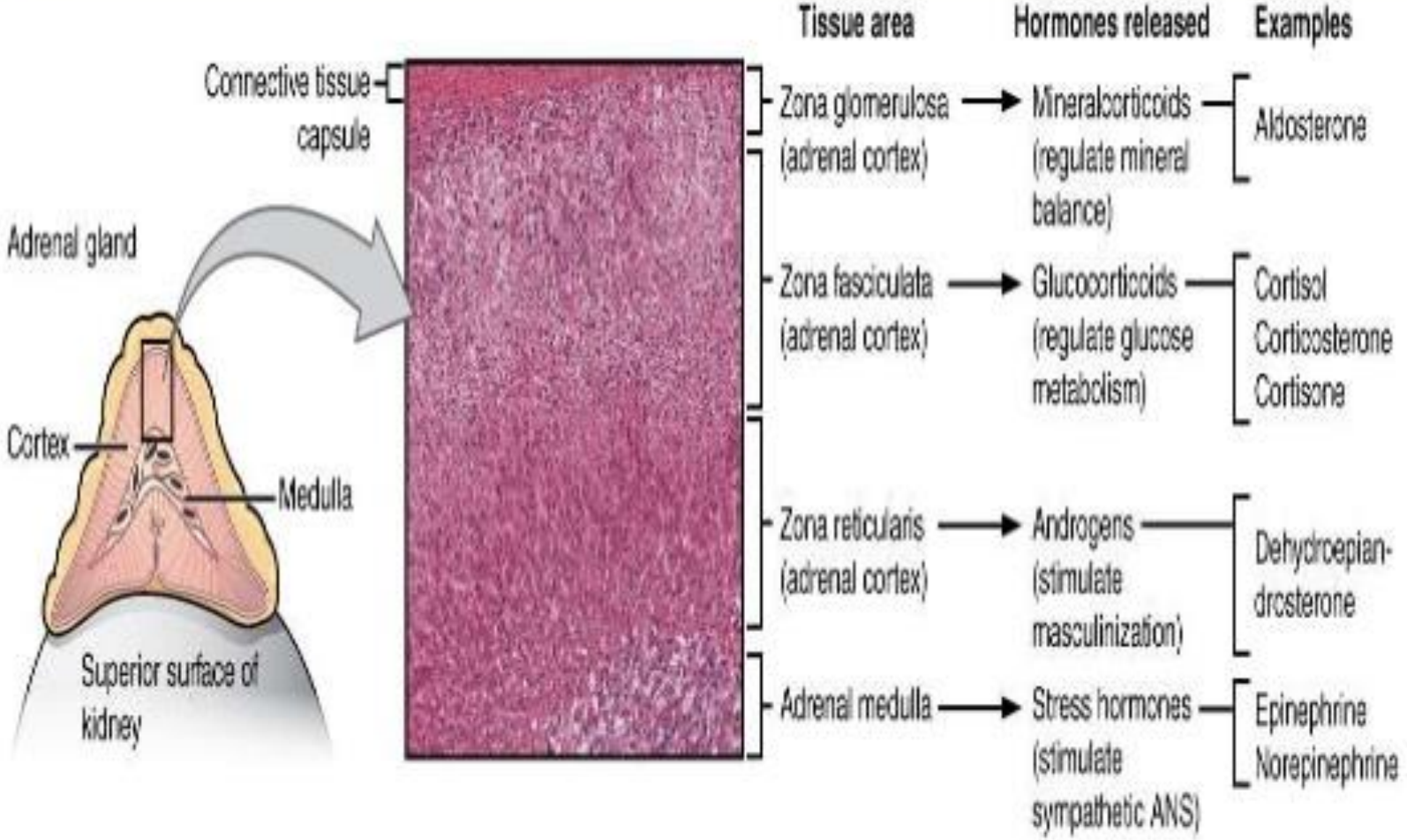
- Cortex and medulla arises from different blastomeric layers
- Adrenal cortex arise from mesodermal cells
- Adrenal medulla arise from neuroectodermal cells, which migrate to cortex from neural crest

HISTOLOGY

- **Adrenal cortex** : Arranged in zonal configuration
  - Outer *zona glomerulosa* = small, compact cells
  - Central *zona fasciculata* = larger, lipid-rich cells arranged in radial columns
  - Inner *zona reticularis* = compact & pigmented cells

- **Adrenal medulla**
  - Thin layers of large chromaffin cells

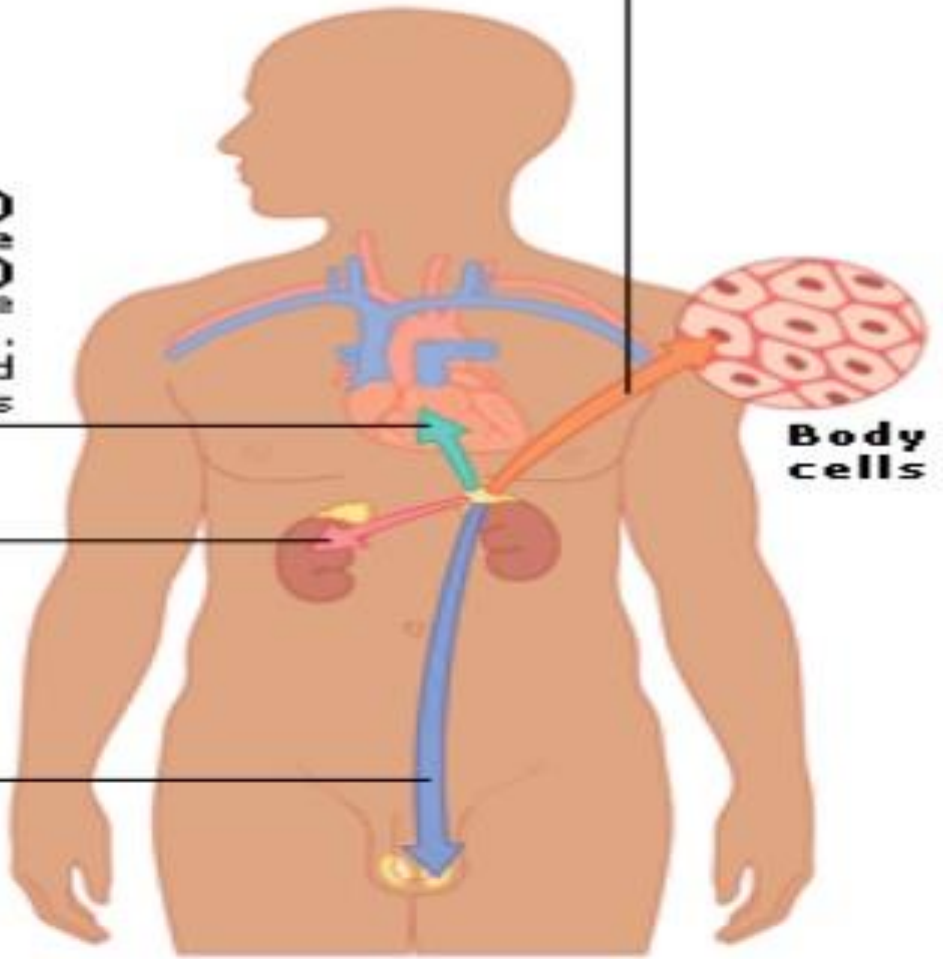
FUNCTION


Connective tissue capsule
Adrenal gland
Cortex
Medulla
Superior surface of kidney
Tissue area
Hormones released
Examples
Zona glomerulosa (adrenal cortex)
Mineralocorticoids (regulate mineral balance)
Aldosterone
Zona fasciculata (adrenal cortex)
Glucocorticoids (regulate glucose metabolism)
Cortisol
Corticosterone
Cortisone
Zona reticularis (adrenal cortex)
Androgens (stimulate masculinization)
Dehydroepian-drosterone
Adrenal medulla
Stress hormones (stimulate sympathetic ANS)
Epinephrine
Norepinephrine

FUNCTION

Cortisol
Cortisol helps the body to adapt to physical and emotional stress by boosting blood glucose levels

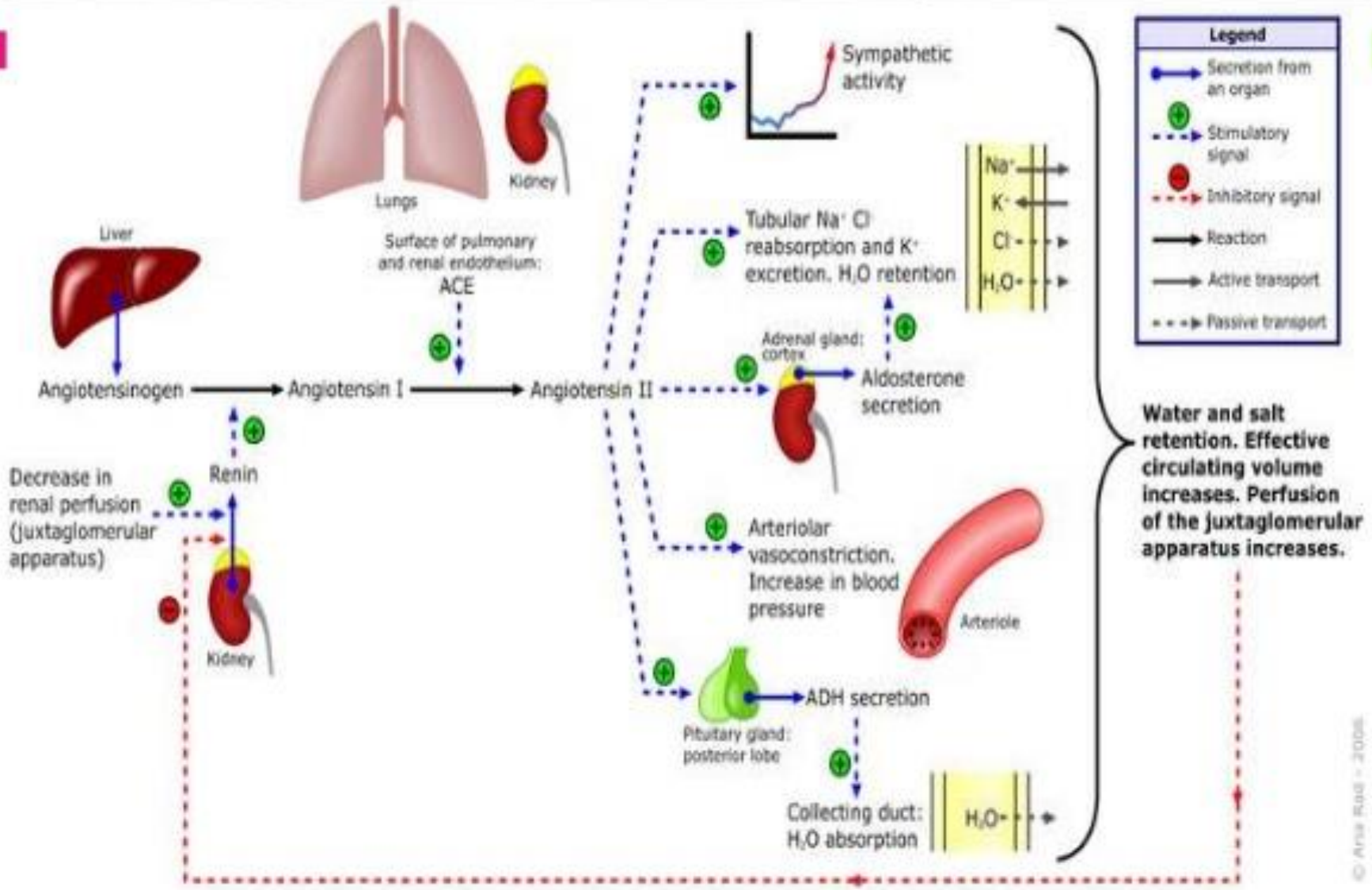
Epinephrine (adrenaline) and norepinephrine (noradrenaline)
These hormones trigger the "fight or flight" response. They increase heart rate and blood flow to the muscles



Aldosterone
This hormone acts on the kidneys to help regulate the excretion of salt to maintain blood pressure

Sex hormones
Adrenal androgens promote the development of secondary male sexual characteristics

Renin-angiotensin-aldosterone system



DISORDERS OF ADRENAL CORTEX

- Incidentaloma
- Primary Hyperaldosteronism
- Cushing's Syndrome
- Adrenocortical Carcinoma
- Congenital Adrenal Hyperplasia
- Adrenal Insufficiency

INCIDENTALOMA

- DEFINITION: a clinically unapparent mass detected incidentally by imaging studies conducted for other reasons.
- INCIDENCE : 1.4%-8.7% , increase with age
- DIAGNOSIS:
- **Hormone evaluation**
  - Morning & midnight plasma cortisol measurements
  - 1mg overnight dexamethasone suppresion test
  - 24 h urinary cortisol excretion
  - 12/24h urinary excretion of metanephrines / plasma-free metanephrines
  - Serum K+, plasma aldosterone & plasma renin activity
  - Serum DHEAS, testosterone/17-hyroxyestradiol
- CT, MRI
- Adrenal gland biopsy (to confirm metastasis)

TREATMENT

- Non-functioning adrenal tumour > 4cm in diameter and smaller tumours that increase in size over time : surgical resection
- Non-functioning tumour < 4 cm: followed-up after 6,12 and 24 months (imaging & hormonal evaluation)

PRIMARY HYPERALDOSTERONISM (PHA)

- Hypertension , Hypokalemia, Hypersecretion of aldosterone
- Hypertensive patient with hypokalaemic PHA~ 2%
- Hypertensive patient have PHA with normal potassium : 12%

PRIMARY HYPERALDOSTERONISM (PHA)

PATHOLOGY

- Most : unilateral adrenocortical adenoma(Conn's syndrome)
- 20-40%: bilateral micronodular hyperplasia
- Rare : glucocorticoid-suppressible hyperaldosteronism/ adrenocortical carcinoma

CLINICAL FEATURES

- Age : 30-50 years old with female predominance
- Hypertension
- Headache
- Muscle weakness
- Cramps
- Intermittent paralysis
- Polyuria
- Polydypsia
- Nocturia

DIAGNOSIS

- Biochemical test
  - Assess potassium and aldosterone level to plasma renin activity ratio
- MRI / CT scan
  - To distinguish unilateral from bilateral disease
  - Conn's adenomas usually measure 1-2cm

- Selective adrenal vein catheterization

✓Samples are obtained from vena cava and both veins

✓Aldosterone to cortisol ratio (ACR) is determined in each sample

✓ACR difference on one side indicate unilateral

TREATMENT

- 1ST line therapy for PHA with bilateral hyperplasia: medical treatment- spironolactone
- Most cases : supplemental antihypertensive medication
- Unilateral laparoscopic adrenalectomy: clear evidence of unilateral/asymmetrical bilateral disease
- Subtotal resection :typical Conn's adenoma

CUSHING'S SYNDROME

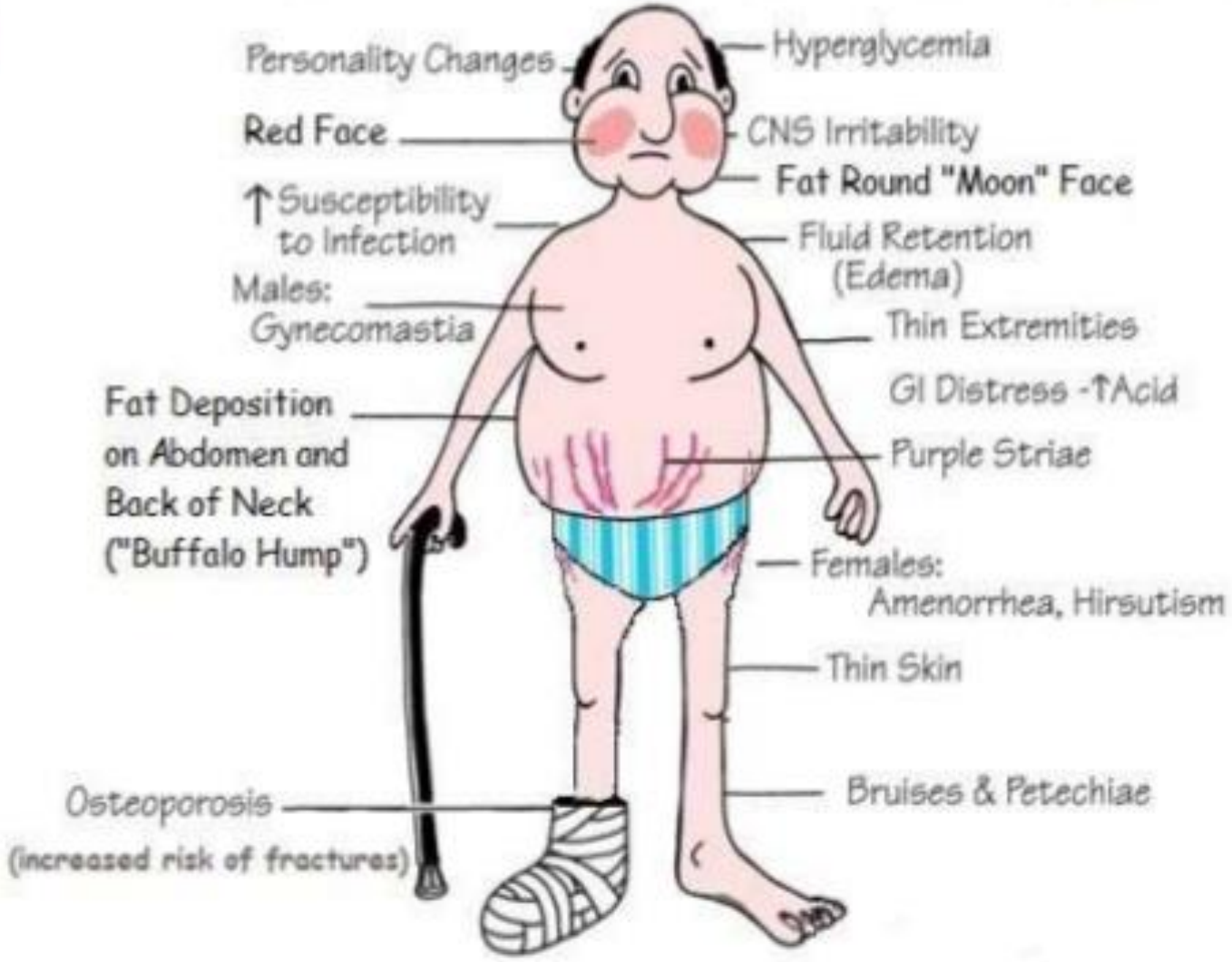
- Hypersecretion of cortisol caused by endogenous production/excessive use of corticosteroids

ACTH-dependent	ACTH-independent
• 85%:Cushing's disease resulting from a pituitary adenoma • Ectopic ACTH-producing tumors • CRH-producing tumors (Medullary thyroid CA, neuroendocrine pancreatic tumor)	• 15%: Unilateral adrenocortical adenoma. • Rare : adrenocortical carcinoma, bilateral micro/macronodular hyperplasia

CLINICAL SYMPTOMS

- Weight gain / central obesity
- Diabetes
- Hirsutism
- Hypertension
- Skin changes (abd striae, facial plethora, ecchymosis, acne)
- Muscle weakness
- Depression / mania
- Osteoporosis
- Hypokalemia

Cushing's Disease or Syndrome Symptoms



DIAGNOSIS

- Morning and midnight plasma cortisol levels are elevated
- Dexamethasone fails to suppress 24-hour urinary cortisol excretion.
- Serum ACTH level

Elevated ACTH level

Performed MRI of pituitary gland

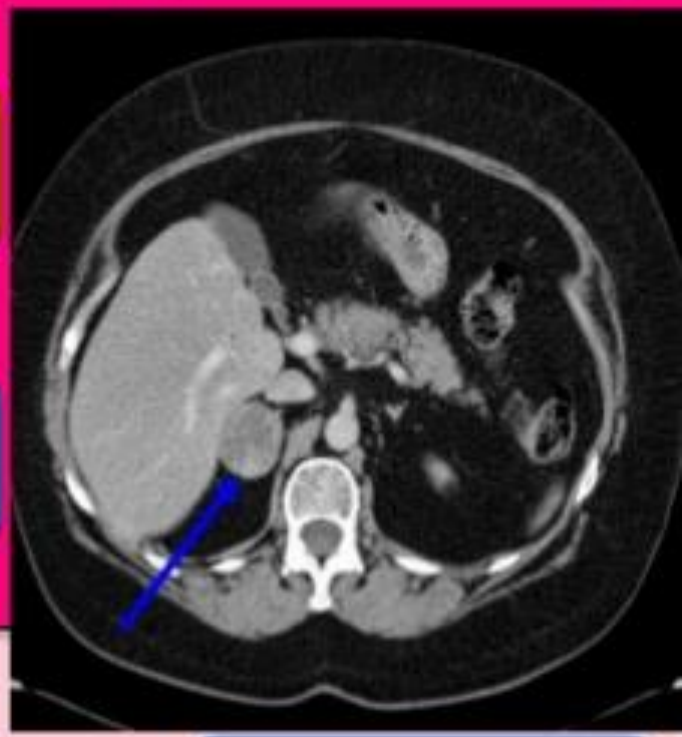
MRI positive

Detect pituitary tumor

MRI negative

CT scan of chest and abdomen

Detect ectopic cortisol producing tumor



Low ACTH level

Performed CT / MRI scan of adrenal gland

Assess adrenal gland

TREATMENT

- Medical therapy with metyrapone or ketoconazole used in patients with severe hypercortisolism or if surgery is not possible.

TYPES OF CUSHING'S SYNDROME	TREATMENT
ACTH-producing pituitary tumours	Trans-sphenoidal resection or radiotherapy.
Ectopic ACTH	Resection
Unilateral adenoma	Adrenalectomy
Bilateral ACTH-independent disease Ectopic ACTH- dependent disease & an irresectable/unlocalised primary tumour	Bilateral adrenalectomy

MANAGEMENT

PRE-OPERATIVE

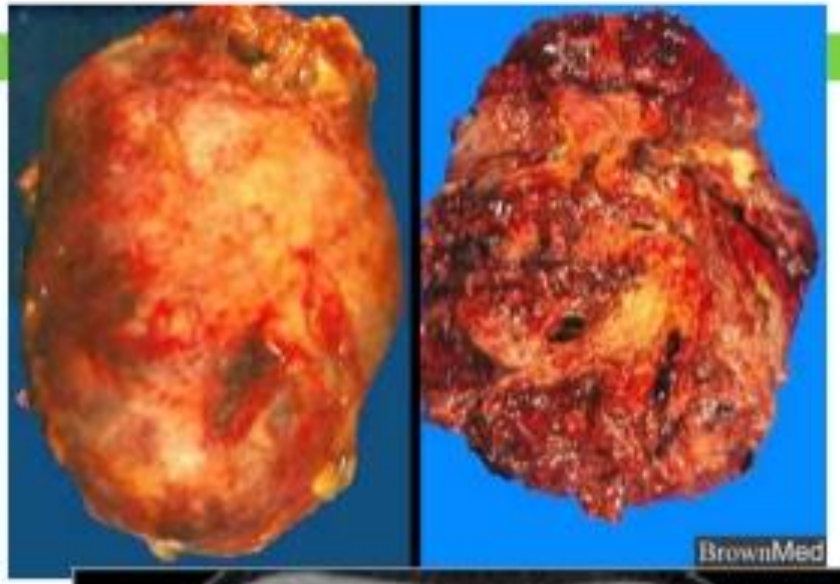
1. Prophylactic anti-coagulation and the use of prophylactic antibiotics are essential
2. Cushing-associated diseases (diabetes, hypertension) must be controlled by medical theraphy

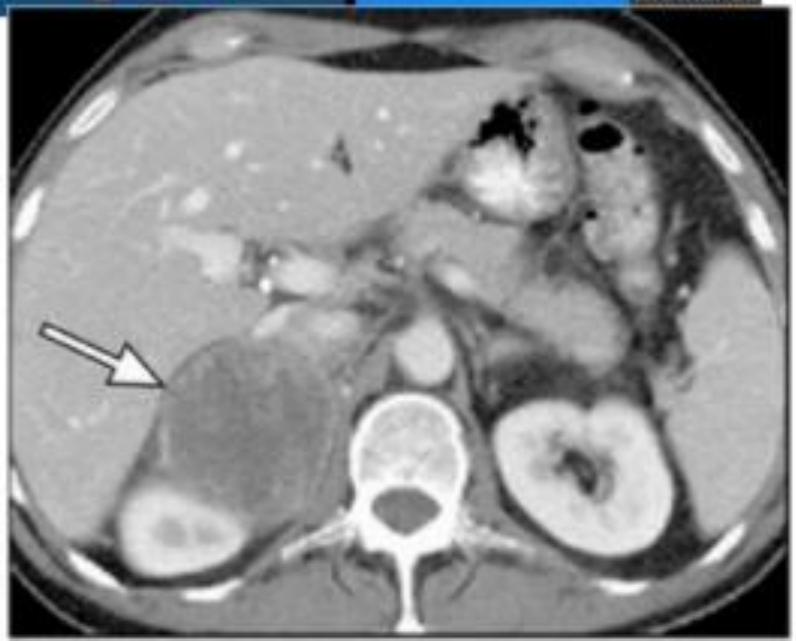
POST-OPERATIVE

1. Supplemental cortisol should be given
   - 15 mg h–1 is required parenterally for the first 12 hours followed by a daily dose of 100 mg for 3 days, which is gradually reduced thereafter.

ADRENOCORTICAL CARCINOMA

- Rare malignancy with an incidence of 1-2 cases in 1mil population per year
- Female predominance (1.5:1)
- Age : 1st peak [childhood], 2nd [4th & 5th decades]





ADRENOCORTICAL CARCINOMA

CLINICAL PRESENTATION

- 60% = cortisol excess (cushing's syndrome)
- Patient with non-functioning tumors
  - Abdominal discomfort
  - Back pain (large tumors)

DIAGNOSIS

- Measurement DHEAS, cortisol, catecholamines
- Dexamethasone suppresion test
- MRI/CT scan
- MRI angiography (exclude tumor thrombus in VC)
- CT scan of lung (distant metastases)

STAGING

- McFarlane classification
- Stage 1 : <5cm
- Stage 2 : >5cm
- Stage 3 : locally invasive tumour
- Stage 4 : tumours with distant metastasis

TREATMENT

- Complete tumor resection (R0)
- *En bloc* resection with removal locally involved organ
- Tumor debulking = control hormone excess
- Tumor thrombus in vena cava = thrombectomy

- **Postoperatively :**
  - Mitotane alone or in combination with etoposide, doxorubicin & cisplatin
  - Adjuvant radiotherapy = reduce local recurrence
  - Restaging for every 3 months = prevent relapse

CONGENITAL ADRENAL HYPERPLASIA (ADRENOGENITAL SYNDROME)

- Virilisation and adrenal insufficiency in children
- Autosomal recessive disorder caused by enzymatic defect in the synthetic pathway of cortisol & other steroid from cholesterol
- 95% : 21-hydroxylase deficiency
- **Common signs :** hypertension & short stature
- **Treatment :**
  - replacement cortisol + fludrocortisone
  - Large hypoplastic adrenals = remove (if symptomatic)

ADRENAL INSUFFICIENCY

- DISEASES ASSOCIATED : polyglandular autoimmune syndroem, TB, after bilateral adrenalectomy, haemorrhage,metastases,etc

Primary
- Loss of function of adrenal cortex (90% adrenal cortex destroyed)

Secondary
- Deficiency of pituitary ACTH secretion

Tertiary
- Loss of hypothalamic CRH secretion
- Therapeutic glucocorticoid administration, brain tumor/ irradiation

ADRENAL INSUFFICIENCY

Acute Adrenal Insufficiency	Chronic Adrenal Insufficiency
• Presents as shock in combination with fever, nausea, vomiting, abdominal pain, hypoglycemia, electrolyte imbalance • Waterhouse-Friderichsen syndrome (bilateral adrenal infarction associated with meningococcal sepsis)	• Presents as anorexia, weakness and nausea • Hyperpigmentation of skin and oral mucosa • Hypotension, hyponatremia, hyperkalemia, hypoglycemia

TREATMENT

- Immediate treatment must be started if patient displays features of adrenal insufficiency.
- IV hydrocortisone 100mg every 6hrs, 3L of saline given in 6hrs under careful cardiac monitoring.

- Chronic adrenal insufficiency = replacement therapy with daily oral hydrocortisone (10mg) and fludrocortisone (0.1mg)

DISORDERS OF THE ADRENAL MEDULLA

- PHAEOCHROMOCYTOMA(ADRENAL PARAGANGLIOMA)
- NEUROBLASTOMA
- GANGLIONEUROMA

PHAEOCHROMOCYTOMA

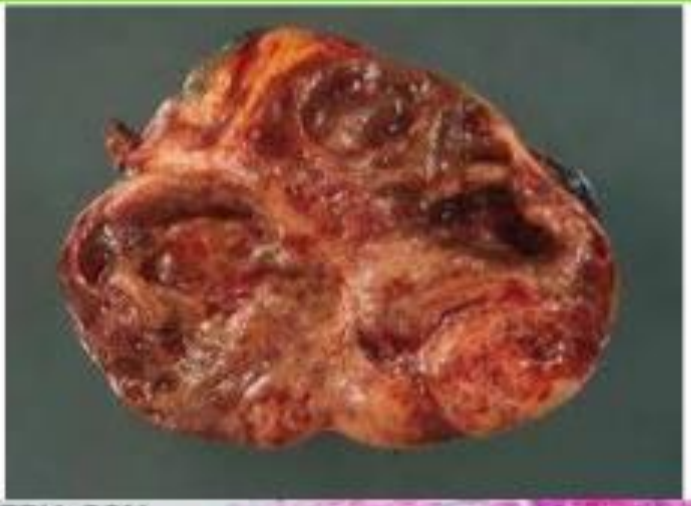
- Tumour of the adrenal medulla, which is derived from chromaffin cells→ catecholamines
- Sporadic occur after 4th decade, hereditary(earlier)
- Known as 10% tumor
  - 10% inherited
  - 10% extra-adrenal
  - 10% malignant
  - 10% bilateral
  - 10% children

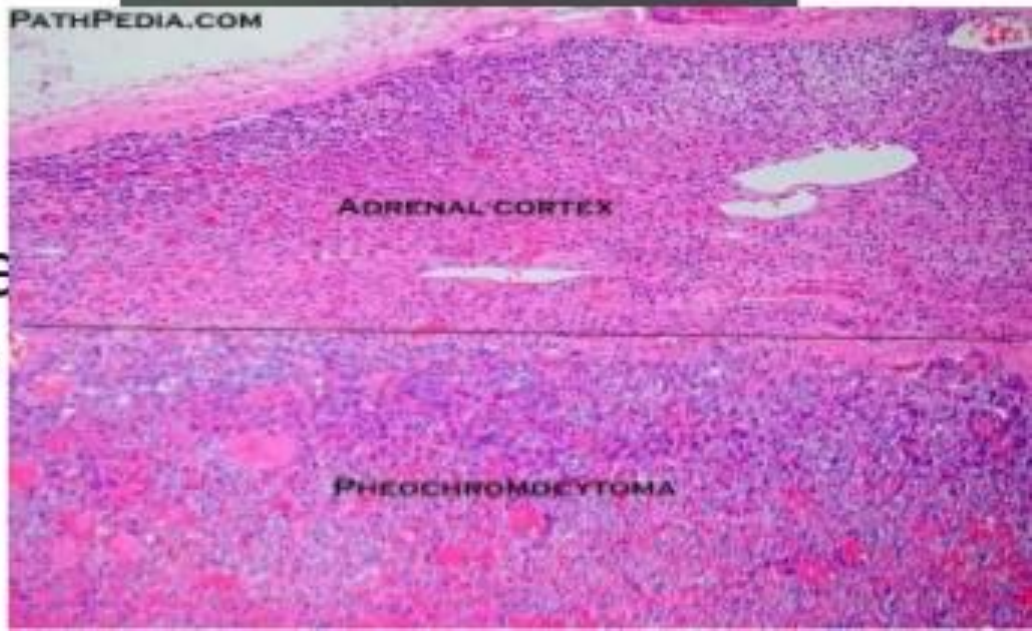
AETIOLOGY

- HEREDITARY PHAEOCHROMOCYTOMAS
- Multiple endocrine neoplasia type 2(MEN 2)
- Familial paraganglioma(PG) syndrome
- Von Hippel-Lindau(VHL) syndrome
- Neurofibromatosis (NF) type 1

PATHOLOGY

- Greyish pink on the cut surface and usually highly vascularized
- Area of haemorrhage & necrosis
- M: polygonal but the configuration varies considerably





CLINICAL FEATURES

Table 49.2 Clinical signs of phaeochromocytoma

Symptoms	Prevalence (%)
Hypertension:	80–90
Paroxysmal	50–60
Continuous	30
Headache	60–90
Sweating	50–70
Palpitation	50–70
Pallor	40–45
Weight loss	20–40
Hyperglycaemia	40
Nausea	20–40
Psychological effects	20–40

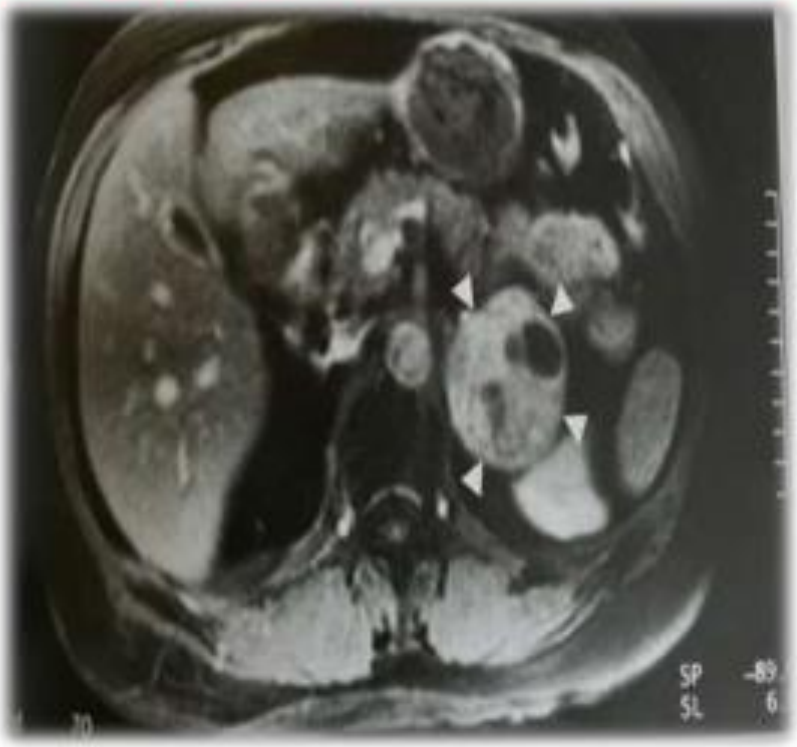
PHAEOCHROMOCYTOMA

- DIAGNOSIS
- Determination of adrenaline, noradrenaline, metanephrine, normetanephrine levels in a 24hour urine collection
- Plasma-free metanephrine& normetanephrine
- MRI-localisation/metastases

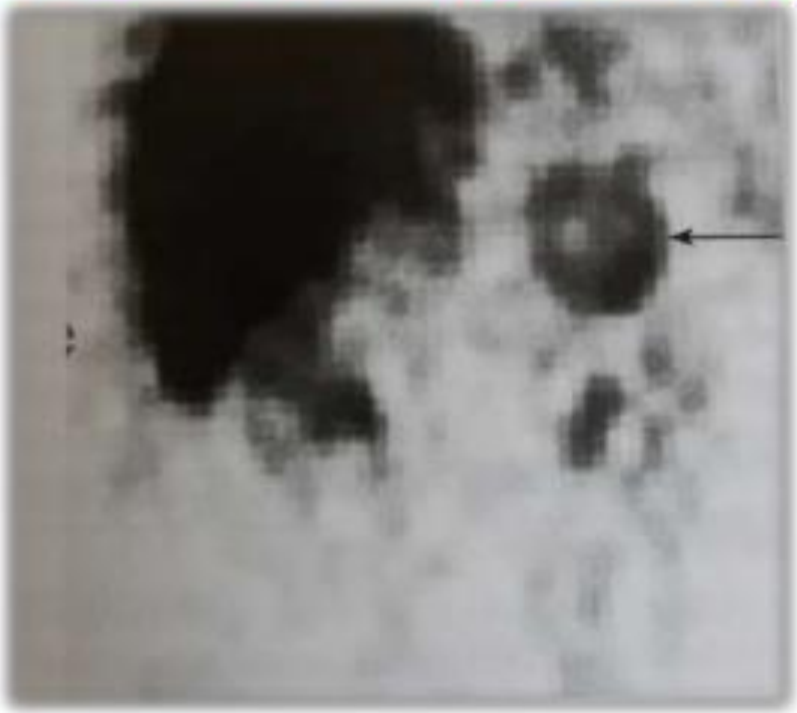
- TREATMENT
- Laparoscopic resection
- >8-10cm/radiological signs of malignancy are detected→ open approach

PHAEOCHROMOCYTOMA

MRI:'swiss cheese' configuration



123I-MIBG (metaiodobenzyl - guanidine) single-photon emission computerised tomography (SPECT)



PHAEOCHROMOCYTOMA

PREOPERATIVE

- α- adrenoreceptor blocker (phenoxybenzamine)
- Additional β- blockade

PEROPERATIVE

- Administration of pressor or vasodilator agents
- Central venous catheter and invasive arterial monitoring

POSTOPERATIVE

- Observed patient 24 hours in ICU
- Lifelong yearly biochemical tests should be performed

MALIGNANT PHAEOCHROMOCYTOMA

- 10% of phaeochromocytoma are malignant
- Higher in paragangliomas
- Metastatsis to lymph nodes, bone and liver

- TREATMENT
- Surgical excision
- Tumor debulking
- Symptomatic treatment by a-blockers
- Mitotane(adjuvant& palliative treatment)
- ^{131}I-MIBG/ combination chemotherapy

PHAEOCHROMOCYTOMA IN PREGNANCY

- Amnion infection syndrome/pre eclampsia
- 1st and 2nd trimester : laparoscopic adrenalectomy after adequate a-blockade
- Risk of miscarriage during surgery is high
- 3rd trimester: elective Caesarean with consecutive adrenalectomy

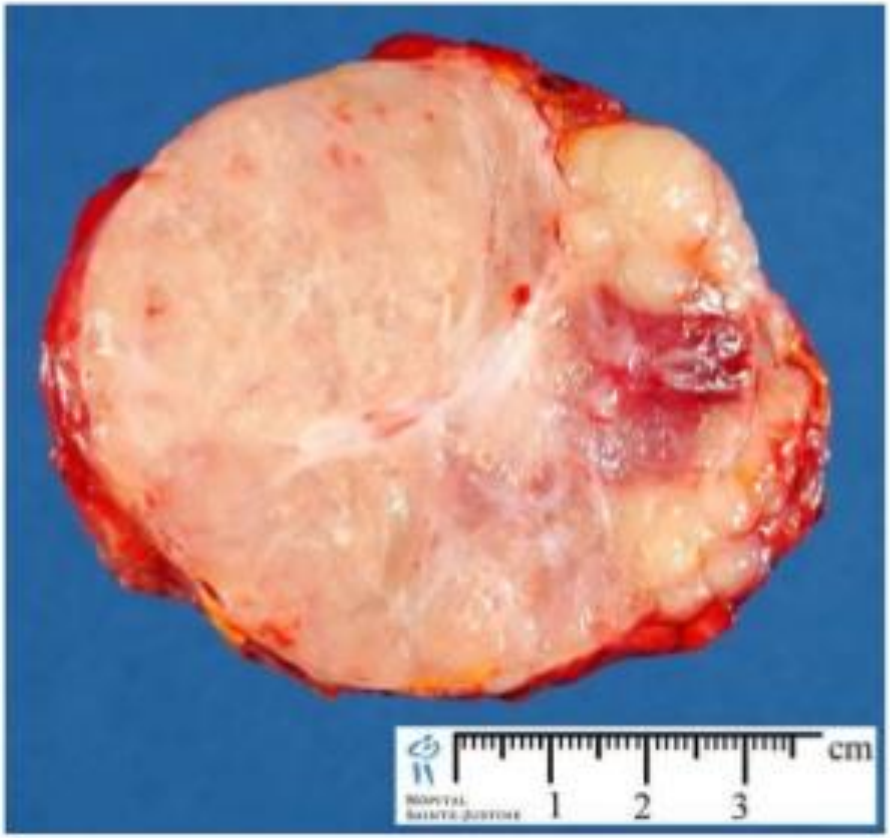
NEUROBLASTOMA

- Malignant tumour that is derived from the sympathetic nervous system in the adrenal medulla (38%)/from any site along the paravertebral sites of abdomen(30%), chest(20%)

- CLINICAL FEATURES
- Newborn infants and young children < 5 years
- Mass in abdomen, neck or chest
- Proptosis/exophthalmus
- Bone pain
- Painless bluish skin metastases
- Weakness or paralyses

PATHOLOGY

- Pale and grey surface
- Encapsulated
- Typical area of calcification
- With increased size of tumour→necrosis and haemorrhage



NEUROBLASTOMA

- DIAGNOSIS
- Biochemical evaluation: urinary excretion(24hour urine) of VMA, HVA, dopamine, noradrenaline
- CT/MRI of the chest& abdomen
- Bone scan,BMA
- MIBG scan

- TREATMENT
- Low-risk:Surgery (the addition of 6–12 w of chemotherapy-optional)
- Intermediate- risk: surgery + multi-agent chemotherapy (carboplatin, cyclophosphamide, etoposide, doxorubicin)
- High-risk patients: high-dose multi-agent chemotherapy followed by surgical resection in responding tumours and myeloablative stem cell rescue.

GANGLIONEUROMA

- DEF: benign adrenal neoplasm that arises from NCT
- Mature sympathetic ganglion cells & Schwann cells in fibrous stroma
- CF: found in all age group(> before 60)
- Occur anywhere along the paravertebral sympathetic plexus & in the adrenal medulla(30%)
- Identified incidentally = CT/MRI for other indications
- TREATMENT: surgical excision, laparoscopic

SURGERY OF THE ADRENAL GLANDS

- LAPAROSCOPIC ADRENALECTOMY:
  - RIGHT ADRENALECTOMY
  - LEFT ADRENALECTOMY
  - RETROPERITONEOSCOPIC ADRENALECTOMY
- OPEN ADRENALECTOMY

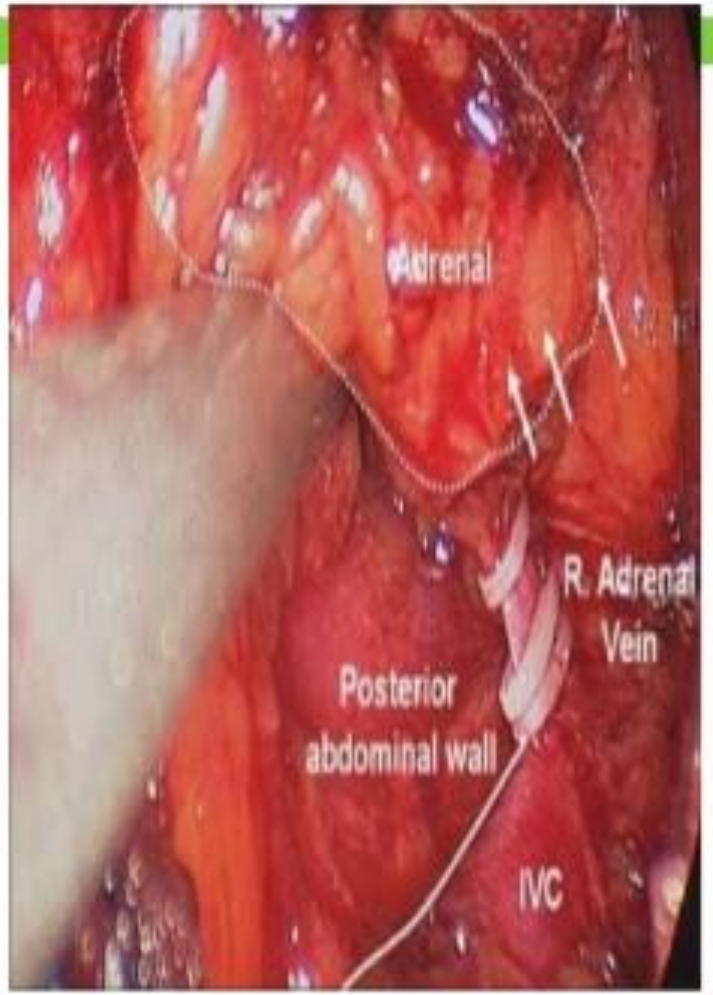
INTRODUCTION

- 1990, laparoscopic/retroperitoneoscopic adrenalectomy→gold standard in the resection of tumor
- Laparoscopic transperitoneal approach: better view of adrenal region
- Small, bilateral tumours/in patients with hereditary tumour syndromes: subtotal resection
- Open approach: radiological signs, dustant metastases, large tumors(>8-10cm)/distinct hormonal pattern suggest malignancy

RIGHT ADRENALECTOMY

- Position: right side up,with table brake
- Dissection start at the level of the periadrenal fat
- Peritoneum is divided 2cm below the edge of liver from medial(IVC) to the lateral abdominal wall
- Flap of peritoneum can be used to retract the liver up & off the adrenal
- Identify the gland and mobilise gently, securing the vein with a clip/using one of the available energy device
- Remove the gland in a plastic catch bag


Instrument port
Camera port
Instrument port


Adrenal
R. Adrenal Vein
Posterior abdominal wall
IVC

LEFT ADRENALECTOMY

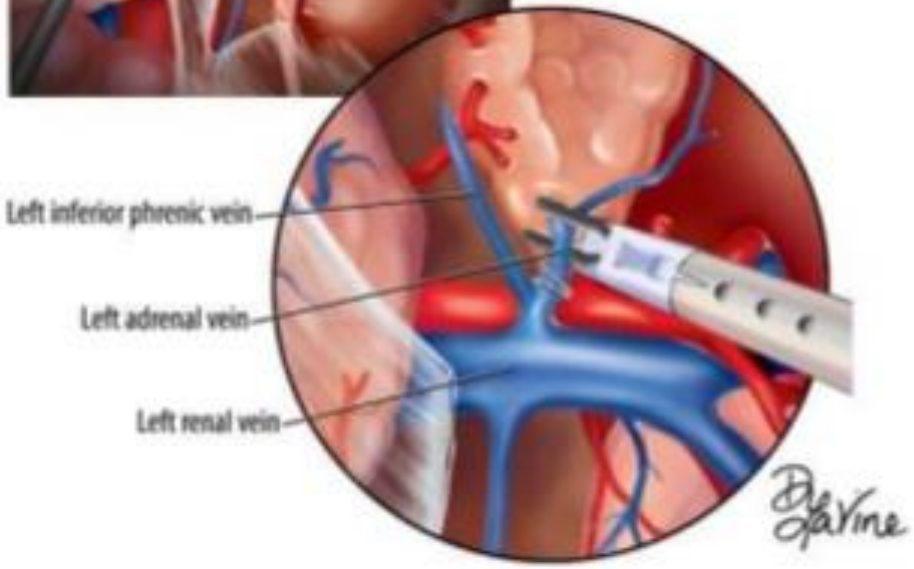
- Position: right side
- Mobilisation of the spleen will displace the pancreatic tail medially
- Incison of Gerota's fascia is followed by identification of the adrenal vein
- The resection is completed by mobilising the adrenal gland at the level of the periadrenal fat
- Remove the gland in a bag

Left Laparoscopic Adrenalectomy

1. Locate the adrenal gland within the perinephric fat.

2. Dissect the gland laterally to medially, exposing the left adrenal vein which drains into the left renal vein. Notice the branch of the left inferior phrenic vein.

3. The vein is clipped twice proximally and once distally before being divided.



RETROPERITONEOSCOPIC ADRENALECTOMY

- 1ST port: distal end of the 12th rib(prone position)
- Digital dissection into the retroperitoneum, Gerota's fascia is displaced ventrally
- RAV is covered by the retrocaval posterior aspect of the adrenal gland
- LAV is located at the medial inferior pole of the adrenal gland
- High inflation pressures allow bloodless dissection effectively tamponading the veins

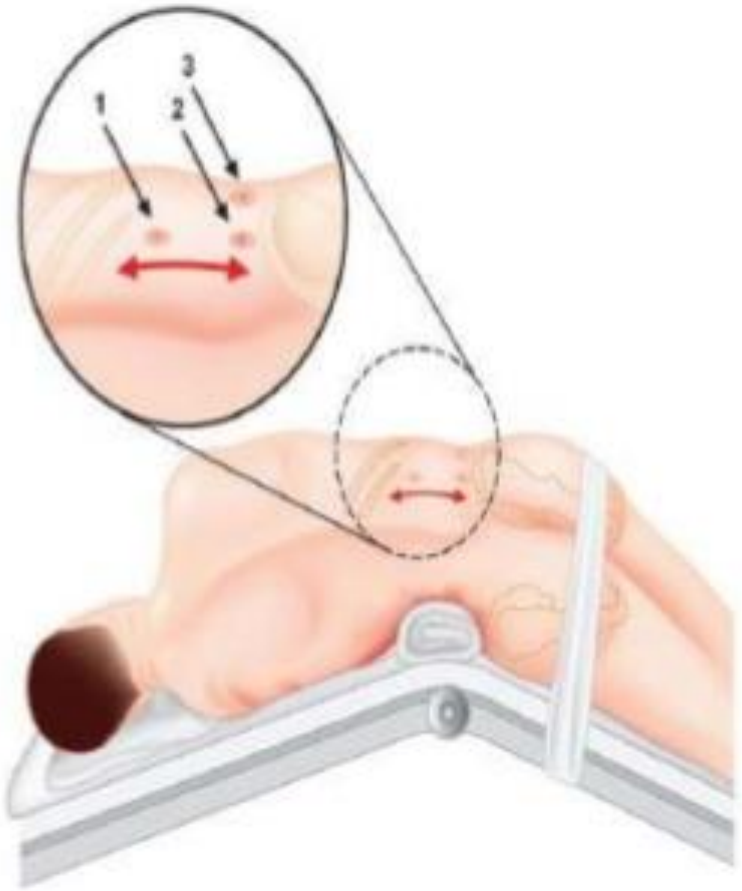


Figure 1 - Lateral position. The patient is placed in a hyperextension position. 1 – first incision is made below the last rib to create a retroperitoneal space; 2 – second port, 1 cm above the iliac crest, where the 0° endoscope enters; 3 – working port located 4 cm anterior to trocar number 2.

OPEN ADRENALECTOMY

- Malignant adrenal tumour suspected
- Rt side: hepatic flexure of the colon is mobilised & the rt liver lobe is cranially retracted to achieve an optimal exposure of the IVC & the adrenal gland
- Lf side: AG can be exposed after mobilisation of the splenic flexure of the colon,through the transverse mesocolon/ the gastrocolic ligament
- Resection of regional lymph node is recommended and should include resection of the tissue between the renal pedicle & diaphragm

REFERENCES

- Bailey & Love's Short Practice of Surgery, 26th Edition,page778-787