

Pheochromocytoma in VHL

James Lee, MD

Edwin K. and Anne C. Weiskopf Associate Professor of Surgery

Chief, Endocrine Surgery

Columbia University Medical Center



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Discover. Educate. Care. Lead.

Disclosures

No relevant disclosures



Overview

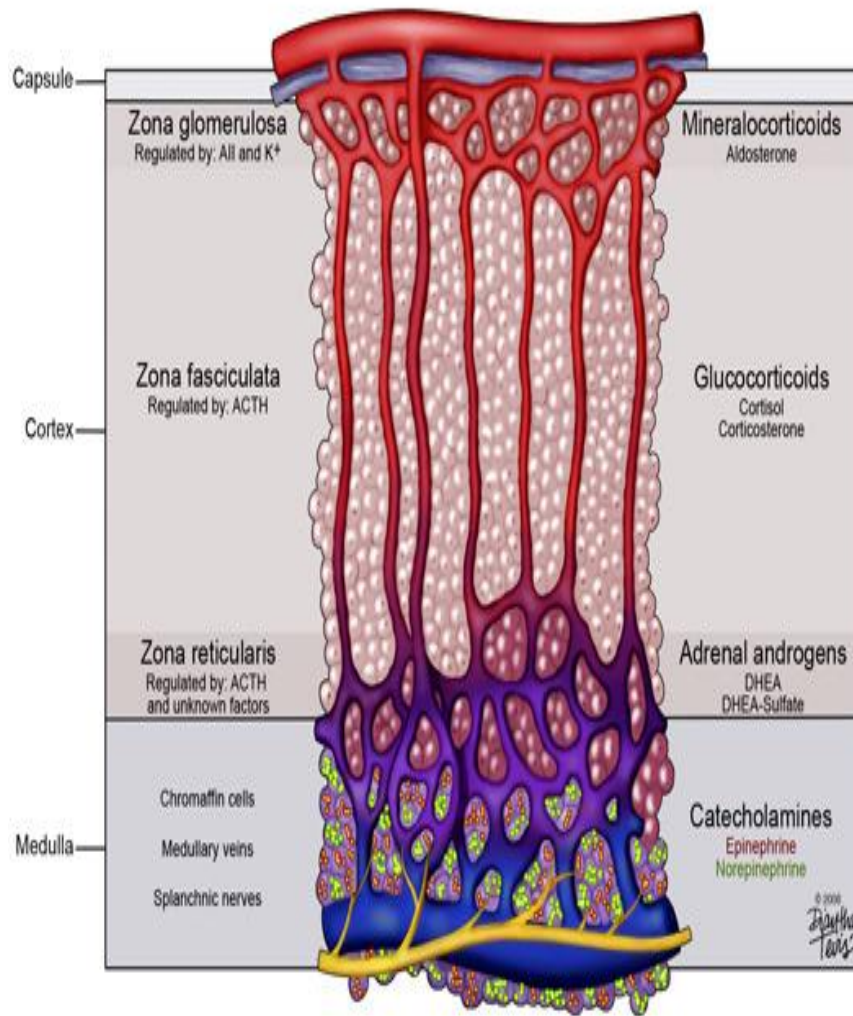
- Background
- Diagnosis
- Treatment



Background



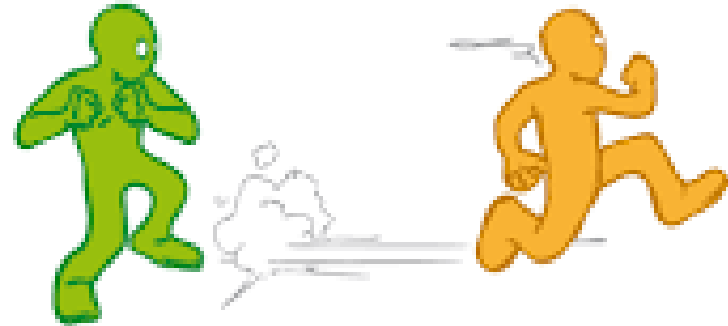
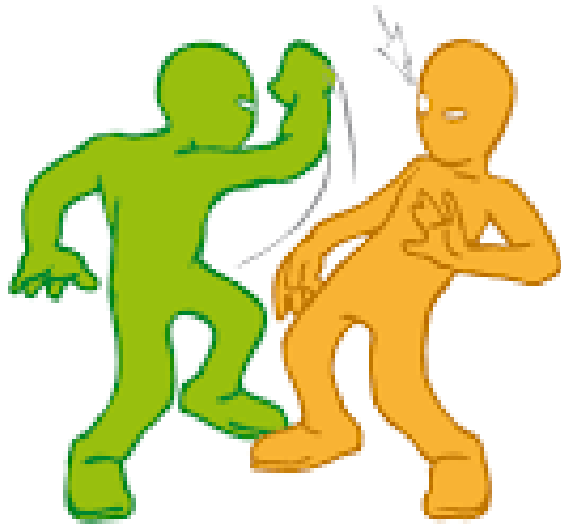




- Cortex:
 - GFR: salt, sweet, sex
- Medulla
 - Chromaffin cells
 - Catecholamines

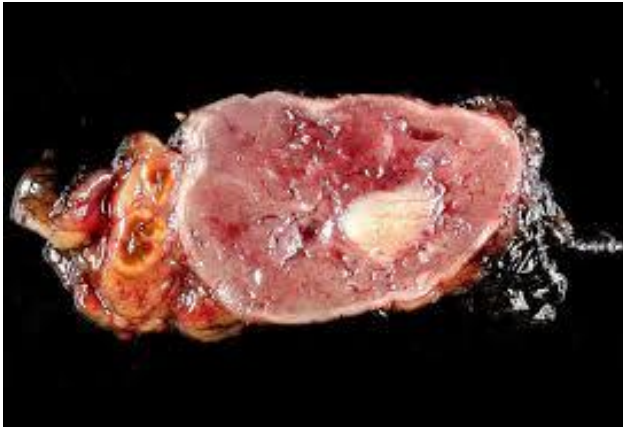


Fight or Flight Hormone

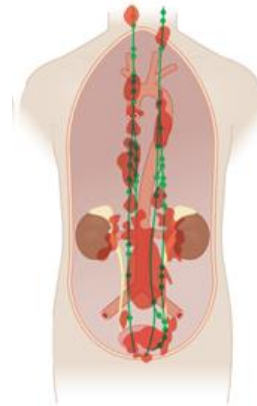


Catecholamine Secreting Tumors

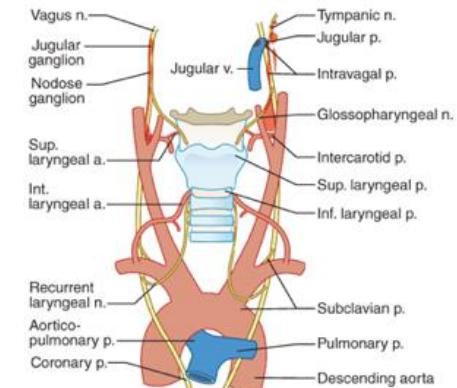
Pheo



Paraganglioma
(Sympathetic)



Paraganglioma
(Parasympathetic)



Symptoms of a Logic Timebomb



High Blood Pressure



Sweating



Palpitations



Headaches

Presentation

- Classic (40%)
 - Palpitation, headache, diaphoresis, hypertension
- Crisis (10%)
 - Multisystem failure, precipitated by procedure
- Screening (10%)
 - MEN2A, MEN2B, VHL, neurofibromatosis
- Incidentaloma (40%)



Epidemiology- all pheos

- Incidence 0.8 per 100,000
- ~1.7% in children with HTN
- Most are sporadic and benign
- 30% have a genetic component

Endocr Pract. 2000;6:249–252

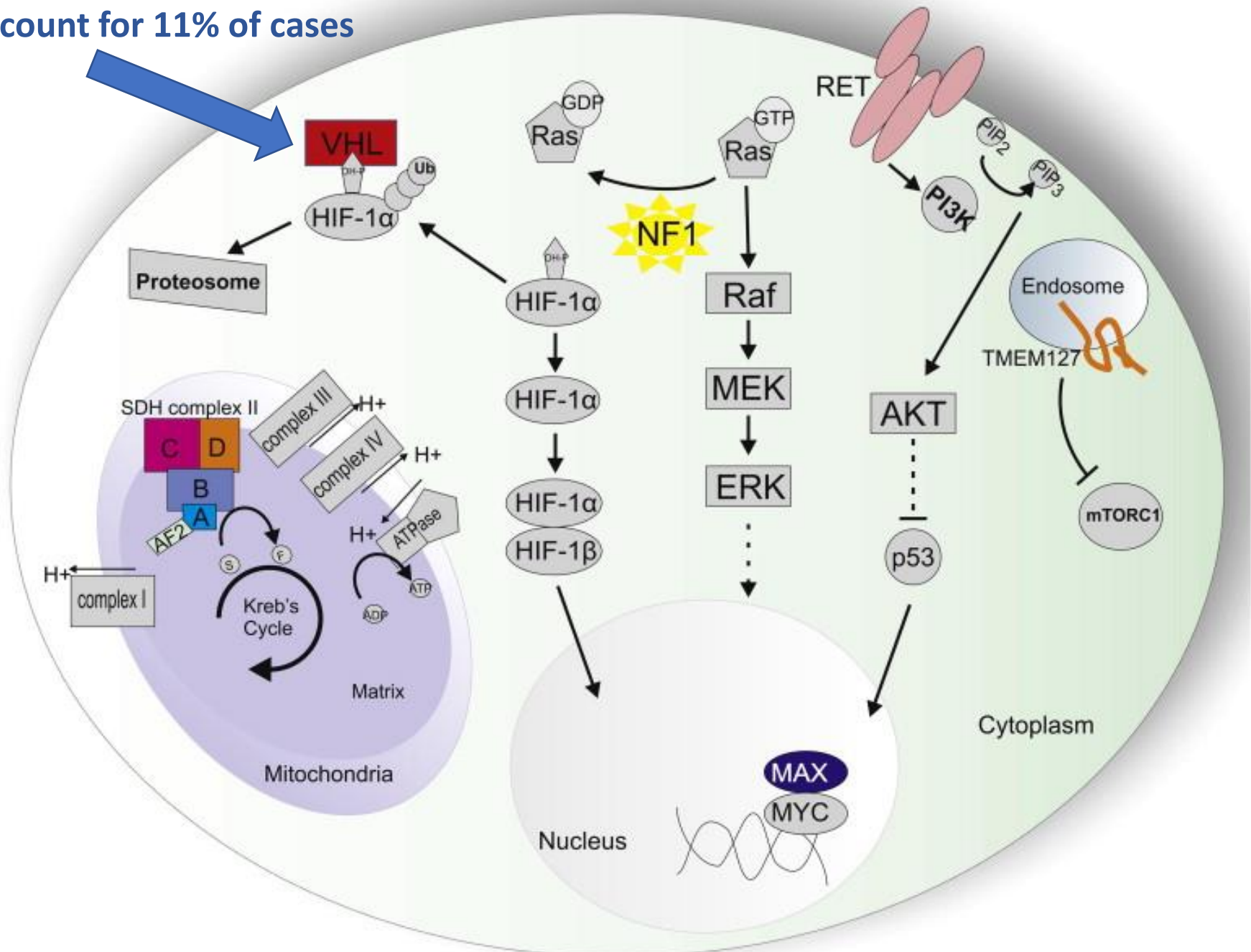
Am J Surg. 2000;179:212–215

J Clin Endocrinol Metab. 2000;85:637–644

JCEM, 2014, 99: 1915-1942.



VHL mutations account for 11% of cases



VHL Subtypes

- VHL Type 1: No pheo
- VHL Type 2: Pheo
 - Type IIA: low risk for renal cell cancer
 - Type IIB: high risk...
 - Type IIC: pheo only



Epidemiology- pheos in VHL

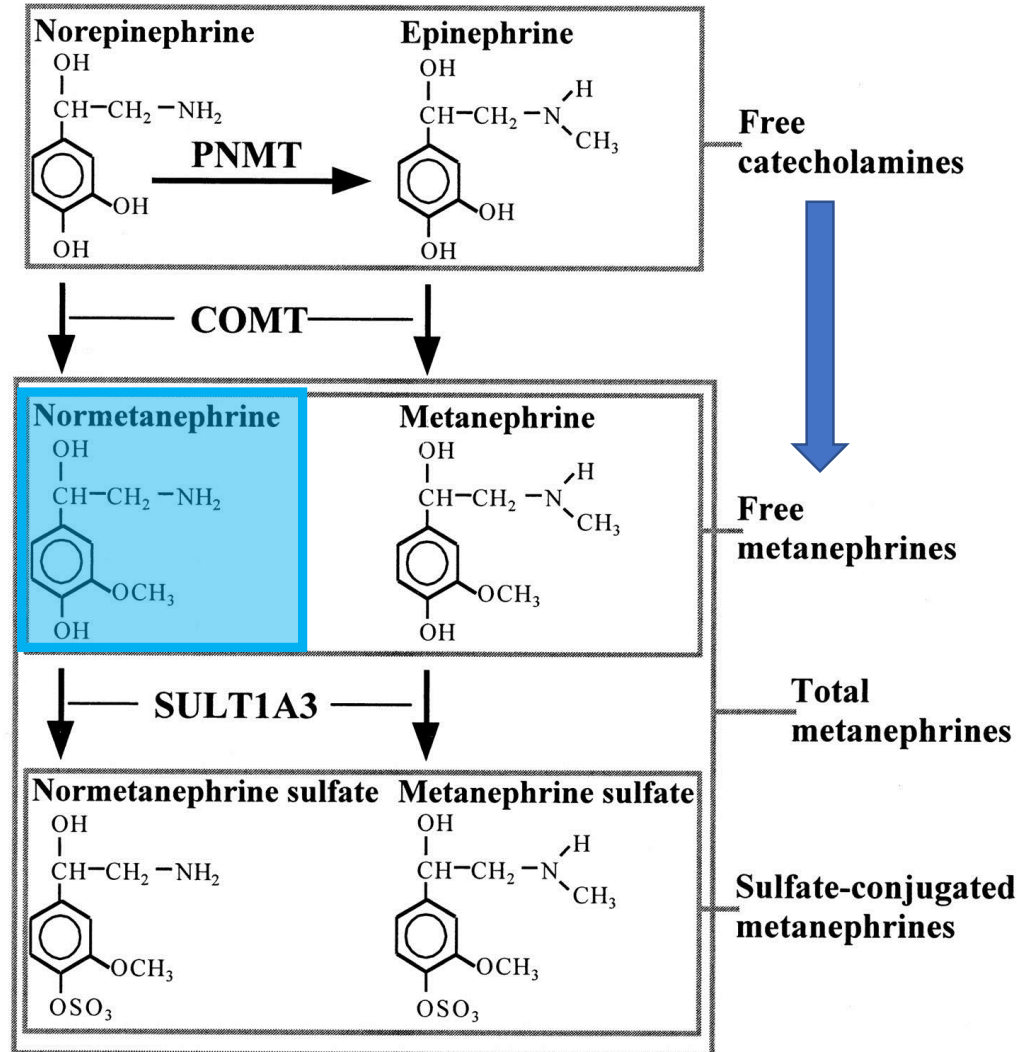
- Typically younger (18% younger than 30)
- Often multiple
- 12% extra-adrenal (paraganglioma)
- 1/3 do NOT produce catecholamines
 - Less likely to be symptomatic



Diagnosis



Metanephrines



Interpreting Metanephrine Levels

- Positive screening test ($> 2\text{-}3\times$ normal)
 - Near 100% positive predictive value for pheo
- Negative screening test ($< 1\times$ normal)
 - Near 100% negative predictive value for pheo
- Borderline ($1\text{-}2\times$ upper limit of normal)
 - 30% positive predictive value for pheo
 - So-called “Subclinical Pheo”



False Positives

- Testing Position
- Age
- Medications

Table 7. Major Medications That May Cause Falsely Elevated Test Results for Plasma and Urinary Metanephrines

	Plasma		Urine	
	NMN	MN	NMN	MN
Acetaminophen ^a	++	—	++	—
Labetalol ^a	—	—	++	++
Sotalol ^a	—	—	++	++
α -Methyldopa ^a	++	—	++	—
Tricyclic antidepressants ^b	++	—	++	—
Buspirone ^a	—	++	—	++
Phenoxybenzamine ^b	++	—	++	—
MAO-inhibitors ^b	++	++	++	++
Sympathomimetics ^b	+	+	+	+
Cocaine ^b	++	+	++	+
Sulphasalazine ^a	++	—	++	—
Levodopa ^c	+	+	++	+

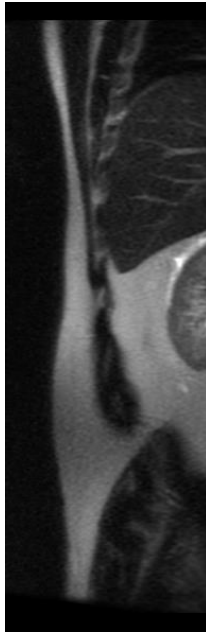


Imaging

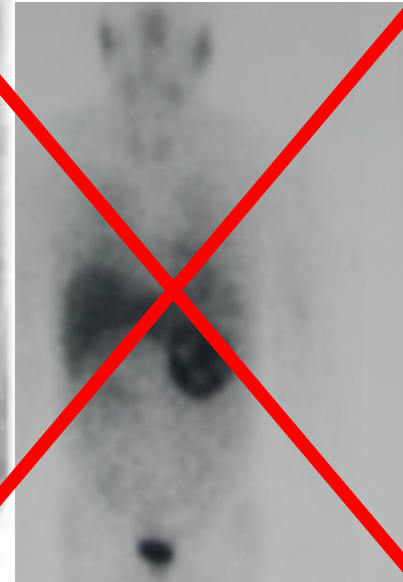
CT Scan



MRI



MIBG



PET



Never biopsy an adrenal tumor until you've ruled out pheo

Screening



Columbia University Medical Center

Von Hippel Lindau Comprehensive Clinical Care Center
Clinical Genetics, BHN Rm 718, 3959 Broadway, NY, NY 10032
212-305-0190, 212-305-0322 FAX

SUGGESTED SCREENING GUIDELINES FOR VHL

- Ages 1-4: Consider plasma and 24 hour urine metanephrines
- Ages 5-15: Plasma metanephrines, abdominal US
- Ages 16 and over: Plasma metanephrines, imaging if abnormal

Treatment



First Resection of a Pheo



- Charlie Mayo
- 1926
- Sister Joachim



Pre-operative Preparation



Alpha Blocker



Beta blocker

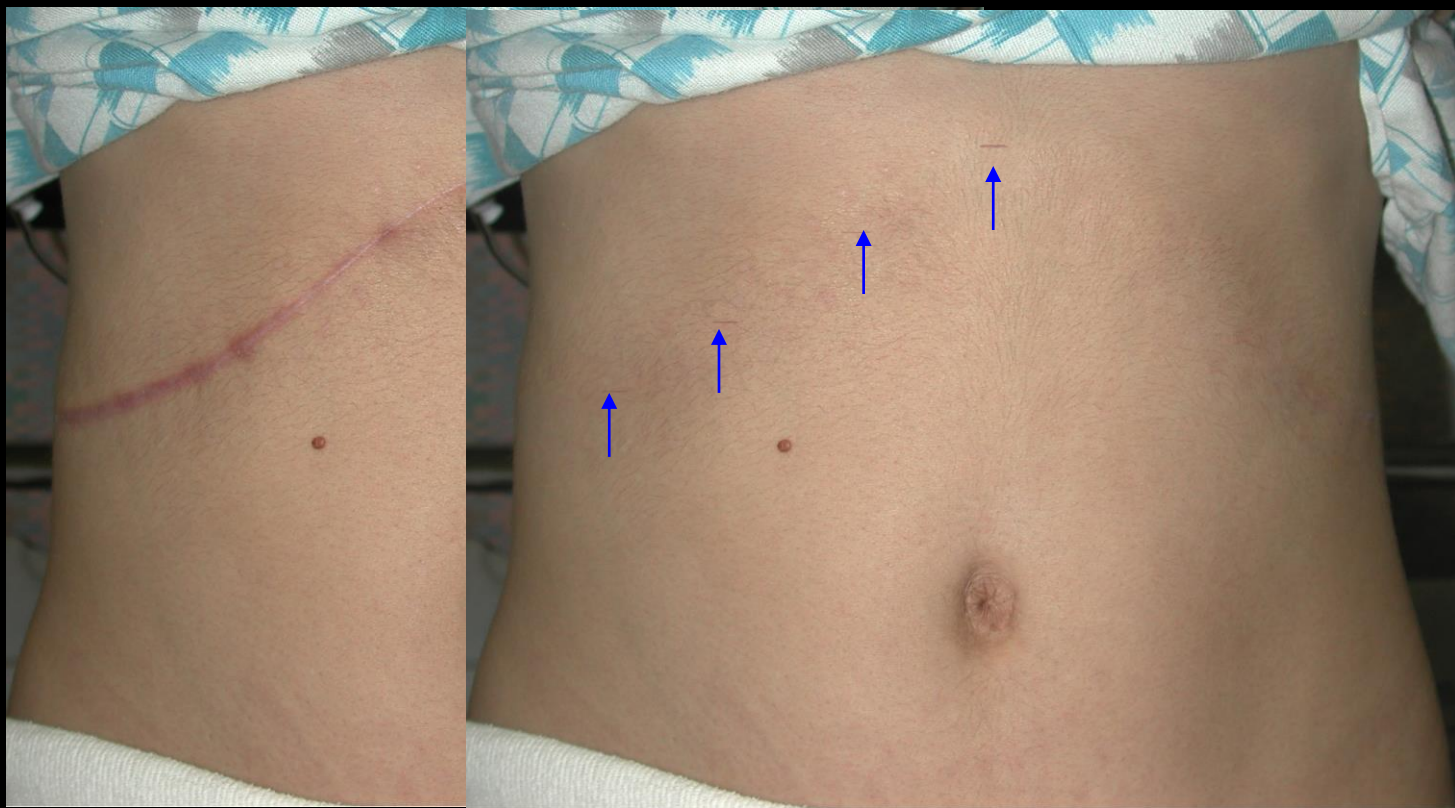
MV04FP83B4P

- Do NOT take a beta blocker until AFTER alpha blockers are started



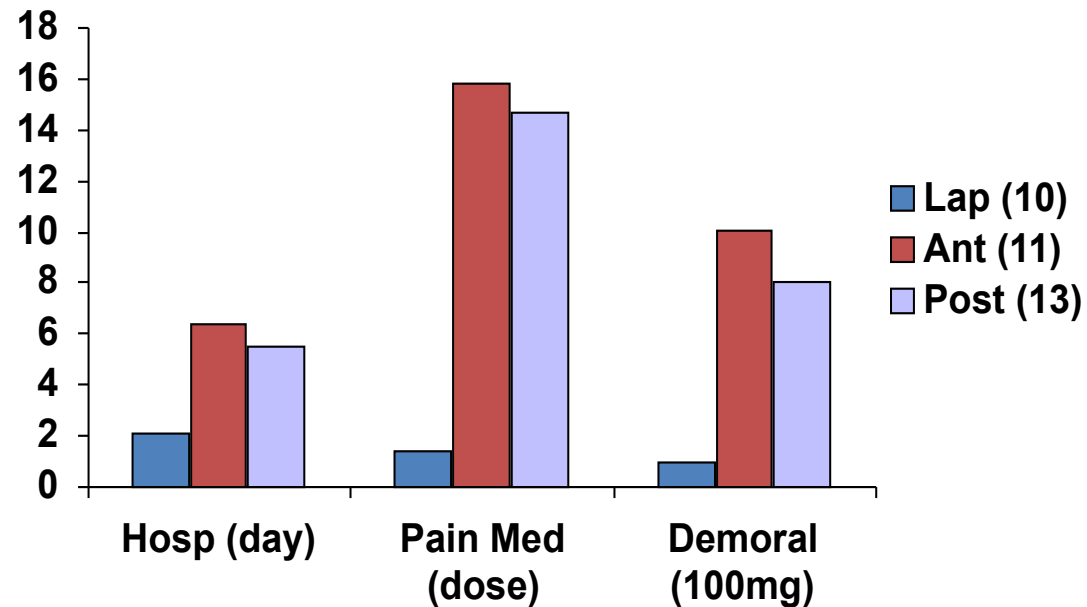
Laparoscopic Adrenalectomy





Better Outcomes With Lap Approach

- Decreased complications, decreased LOS, increased patient satisfaction and comfort



UCSF Database, 4/93-9/04

Park HS, etc al. Arch Surg 144(11):1060-1067

Prinz, RA: Western Surgical, Arch Surg 1995

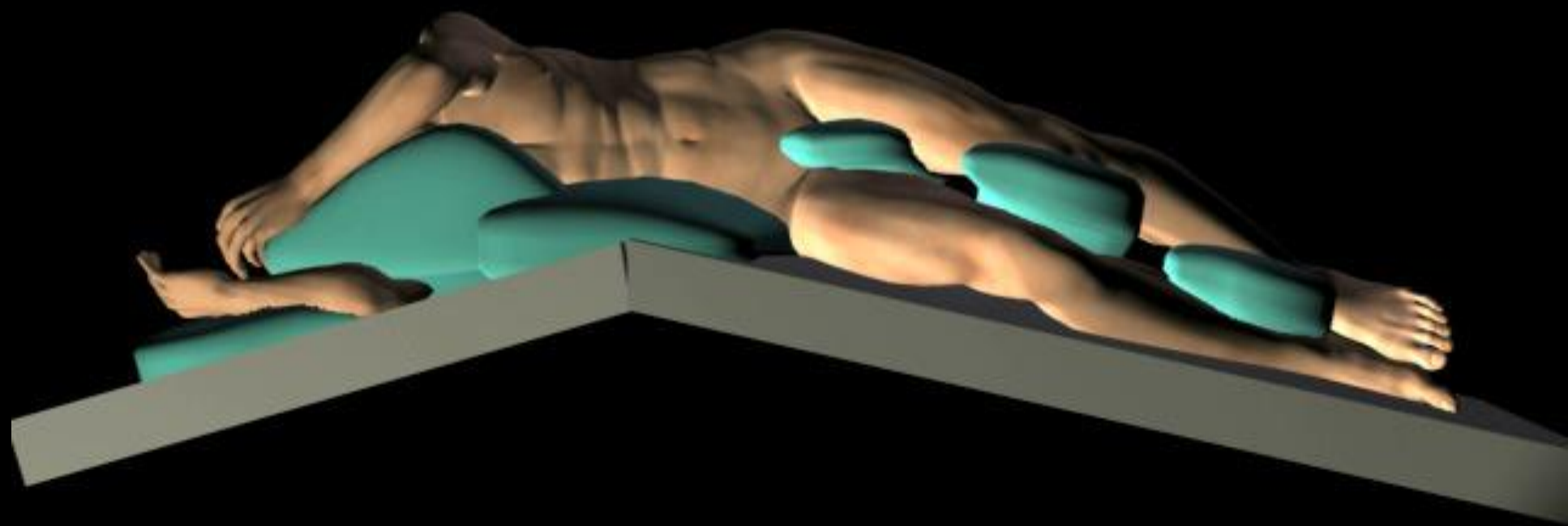


Traditional Approach: Lateral Transabdominal



- More familiar anatomy (peripheral view)
- Diagnostic laparoscopy (mets?)
- Larger working space (tumors >6cm)
- Flexible



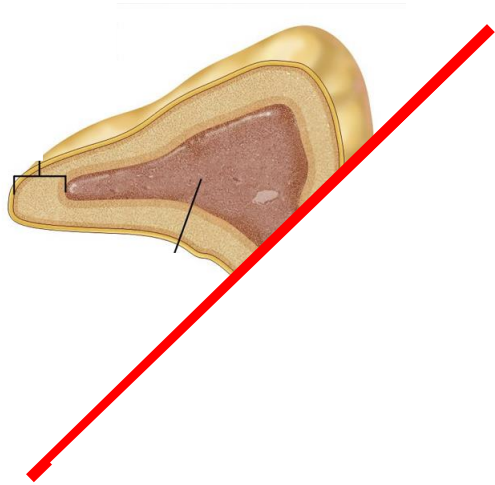


Advantages of Retroperitoneal Approach

- Avoid abdomen in patients with prior abdominal surgery
- More direct approach to adrenal glands
- Fewer complications
- Faster operating time
- Less blood loss
- No need to reposition the patient for bilateral adrenalectomy

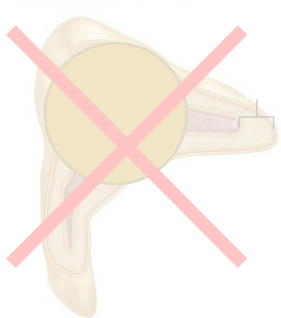


Cortical Sparing Adrenalectomy

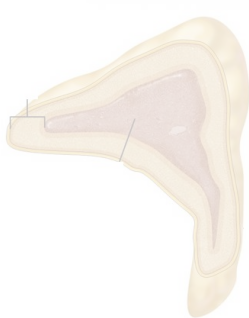


Staged Adrenalectomy

Stage 1

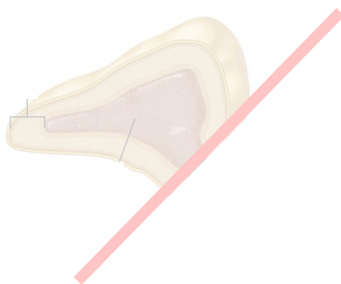
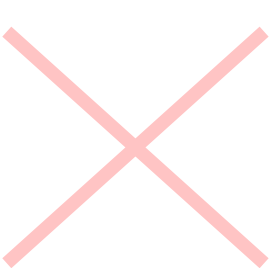


Right

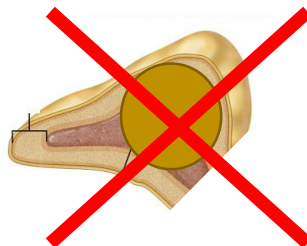
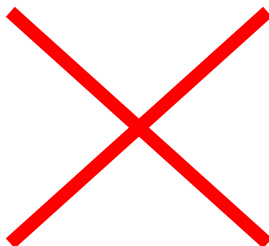


Left

Stage 2



Stage 3



Questions?

