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Kidney and Upper Urinary Tract

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Fall 2008

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Syllabus

- If I show you graphics that are NOT in your syllabus
  - Then they are NOT critical for the test
  - They are to familiarize you with Urologic operative techniques, for their interest rather than for testing purposes
- Radiography will be on test, but not the actual images

Objectives

- Appreciate importance, evaluation, and differential diagnosis of hematuria
- Gain basic understanding of major disease processes of kidney and upper tract
  - Obstruction
  - Calculi
  - Infection
  - Renal masses and cysts

Kidney and Upper Urinary Tract

Clinically-Oriented Lecture

Hematuria

↓

Evaluation and Differential

↓

Case presentations of representative entities

Hematuria

- Definition
  - ≥ 3 RBC / hpf in urinary sediment
  - Dipstick is screening test only
  - Dipstick is 95 % sensitive, but only about ~ 80% specific
  - In population with 10% hematuria, PPV of Dipstick is only 35%
  - Positive Dipstick indicates hematuria ONLY when confirmed by microscopy of ≥ 3 RBC / hpf

Hematuria

- Additional Characterization
  - Gross (grossly visible) or microscopic?
    - Gross more likely significant
  - If gross, is it initial (urethra), terminal (bladder neck or prostate), or total (interior of bladder or upper tract)

Evaluation 1: Examine Urine

- In women, get cath. UA if > 1 squamous cell / hpf (vaginal contamination)
- Is color red but dipstick - ?
  - ➔Consider phenolphthalein, rhodamine B, others
- Is dipstick + but no RBC present?
  - ➔Beware if specific gravity < 1.008 (RBC may have been there, but lysed)
  - ➔Usually false positive test
  - ➔Unfortunately, a common referral

Evaluation 1: Examine Urine

- Is there pyuria or bacteruria?
  - ➔Probable infection (any GU site)
- Is there proteinuria (> 2 + on dipstick), or are there dysmorphic RBCs or RBC casts?
  - ➔Probable glomerulonephritis

Evaluation 2: History and Physical

- Flank Pain
  - → Stone
- Dysuria, bladder irritability
  - → Bladder or prostate infection
- Sickle cell, diabetes
  - → Papillary necrosis
- Family or personal history of calculi, PCKD, other GU / Neph diseases
  - → Possible familial trait

Evaluation 2: History and Physical

- Trauma, or intense physical activity
  - ➔ May be cause of hematuria
- Tobacco use, occupational chemical exposure (aromatic dyes)
  - ➔ Risk for renal and urothelial cancers

Evaluation 2: History and Physical

- Visible blood at urethral meatus
  - → Urethral source
- Fever, CVAT
  - → Pyelonephritis
- Prostate exam
  - → Prostatitis, Prostate cancer
- Pelvic Exam
  - → Urethral, vaginal, or labial lesions

Evaluation 3: Labs and Procedures

- Formal urinalysis with microscopic examination
- Urine culture
  - If infection is DOCUMENTED, then can omit rest of work-up if hematuria clears with antibiotics
- IVU versus KUB + US versus CT
- +/- Urine cytology
- +/- Serum electrolytes and creatinine
- Cystoscopy

Diagnostic Categories

- Infection
- Calculi
- Cancer
- Benign neoplasms / lesions
- Other obstruction
- Trauma / exertional hematuria
- Medical renal disease
- Blood dyscrasia / anticoagulation
- Benign familial hematuria

Infection

- Kidney
  - Pyelonephritis - parenchyma
  - Pyonephrosis - pus in collecting system
  - Renal abscess - pus pocket in parenchyma

Infection

- Bladder
  - Bacterial cystitis
- Prostate
  - Bacterial prostatitis
- Urethra
  - Infectious urethritis

Calculi

- Kidney
  - Obstructive vs. Non-obstructive
  - Simple vs. Staghorn
- Ureter
  - Obstructive vs. Non-obstructive
- Bladder

Cancer

- Kidney
  - Renal cell carcinoma, other
- Upper collecting system
  - Urothelial, other
- Bladder
  - Urothelial, other
- Urethra
  - Squamous cell, other
- Prostate
  - Adenocarcinoma, other

Benign Neoplasms / Lesions

- Kidney
  - Simple cysts
  - Cystic renal diseases
  - Angiomyolipoma, other neoplasms
- Ureter
  - Hemangioma, other
- Bladder
  - Endometrioma, other
- Urethra
  - Condyloma, other

Other Obstructions

- Ureter
  - Ureteropelvic junction (UPJ)
  - Intrinsic strictures
  - Extrinsic obstruction
- Bladder
  - Bladder outlet obstruction (BOO)
    - Benign prostatic hyperplasia (BPH)
    - Other
- Urethra
    - Strictures, other

Trauma / Exertional Hematuria

- Cannot be assumed to be cause

Medical Renal Disease

- Previous Nephrology lecture

Blood Dyscrasia / Anticoagulation

- Cannot be assumed to be cause

Benign Familial Hematuria

- Microscopic only, negative work-up

Kidney, Intra-renal Collecting System, and Ureter

- Topics covered (case presentations)
  - Ureteral obstruction (UPJO)
  - Calculi (ureteral, renal)
  - Infection (pyelonephritis)
  - Cancer (renal cell carcinoma)
  - Renal cystic disease (ADPKD)

Case 1: Ureteropelvic Junction Obstruction (UPJO)

- 24 year old woman
- Long history of intermittent left flank pain, recently worsening
- Not during sleep
- Especially notices after fluid intake, or even more after drinking alcohol
- No significant medical history

Case 1: Ureteropelvic Junction Obstruction (UPJO)

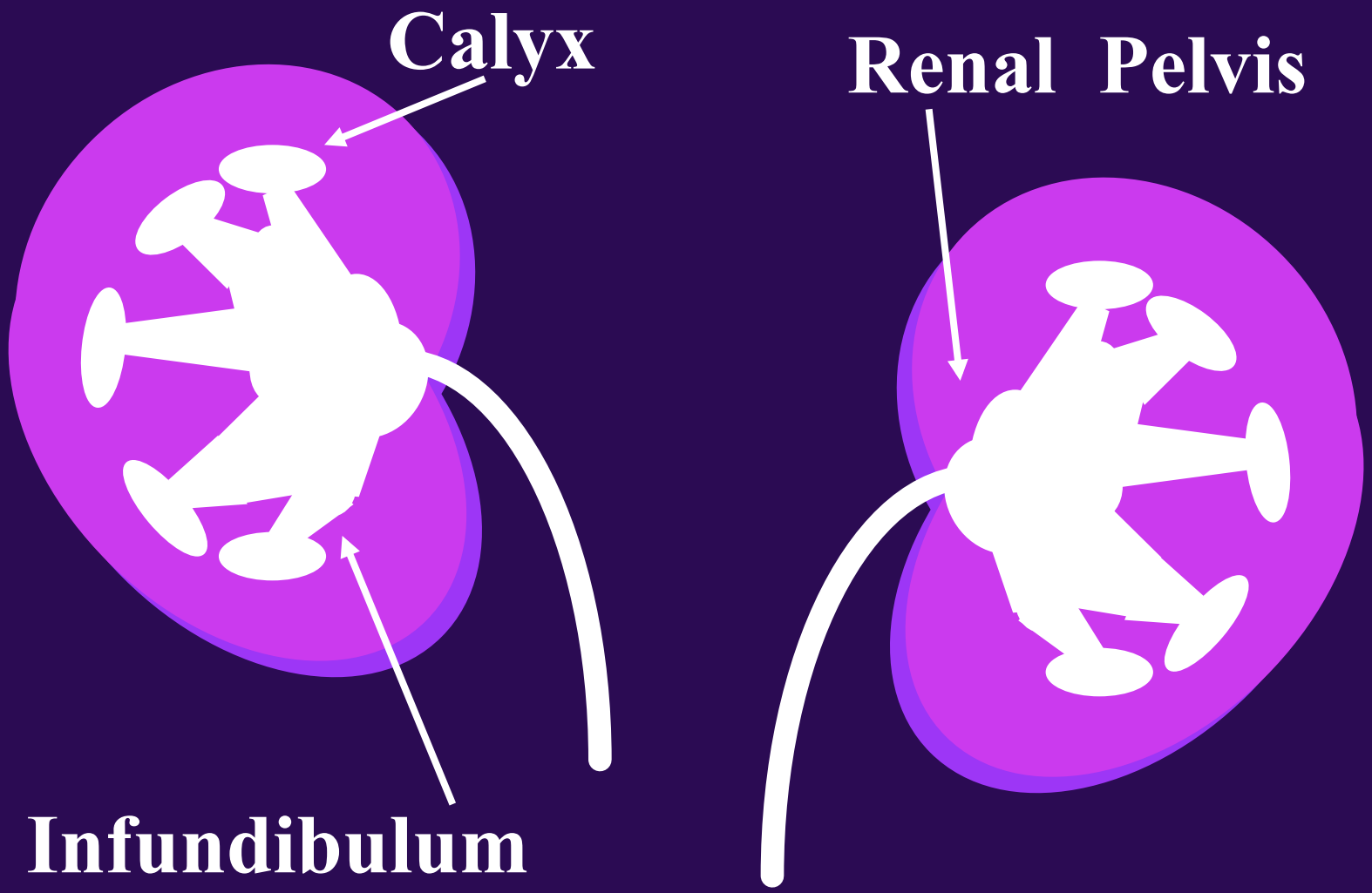
- Young age - diagnosed mostly in children, variable after that
- Intermittent symptoms - typical of adult presentation
- Pain increased with fluid intake
  - Classic for UPJO
  - Flow-dependent obstruction (like a slow but not-yet clogged drain)

Kidney & Upper Urinary Tract: Obstruction

Anatomy of Kidney and Upper Urinary Tract

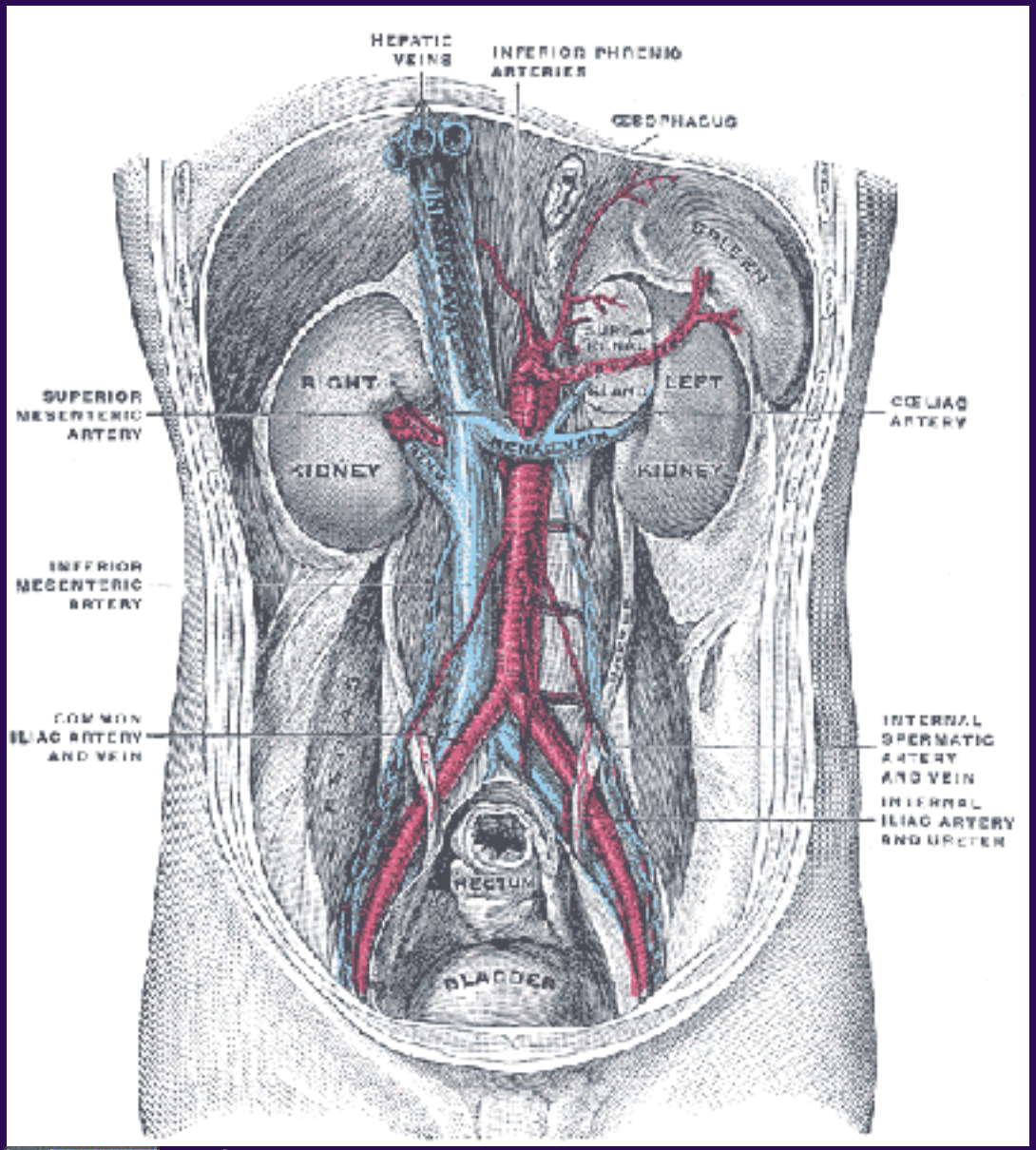
- Paired Kidneys
- Urine from collecting tubules that terminate in papillae drain into:
  - Calyces, that coalesce into
  - Infundibula, which drain into
  - Renal Pelvis
- Travels down ureter (peristalsis)

Kidney & Upper Urinary Tract: Obstruction

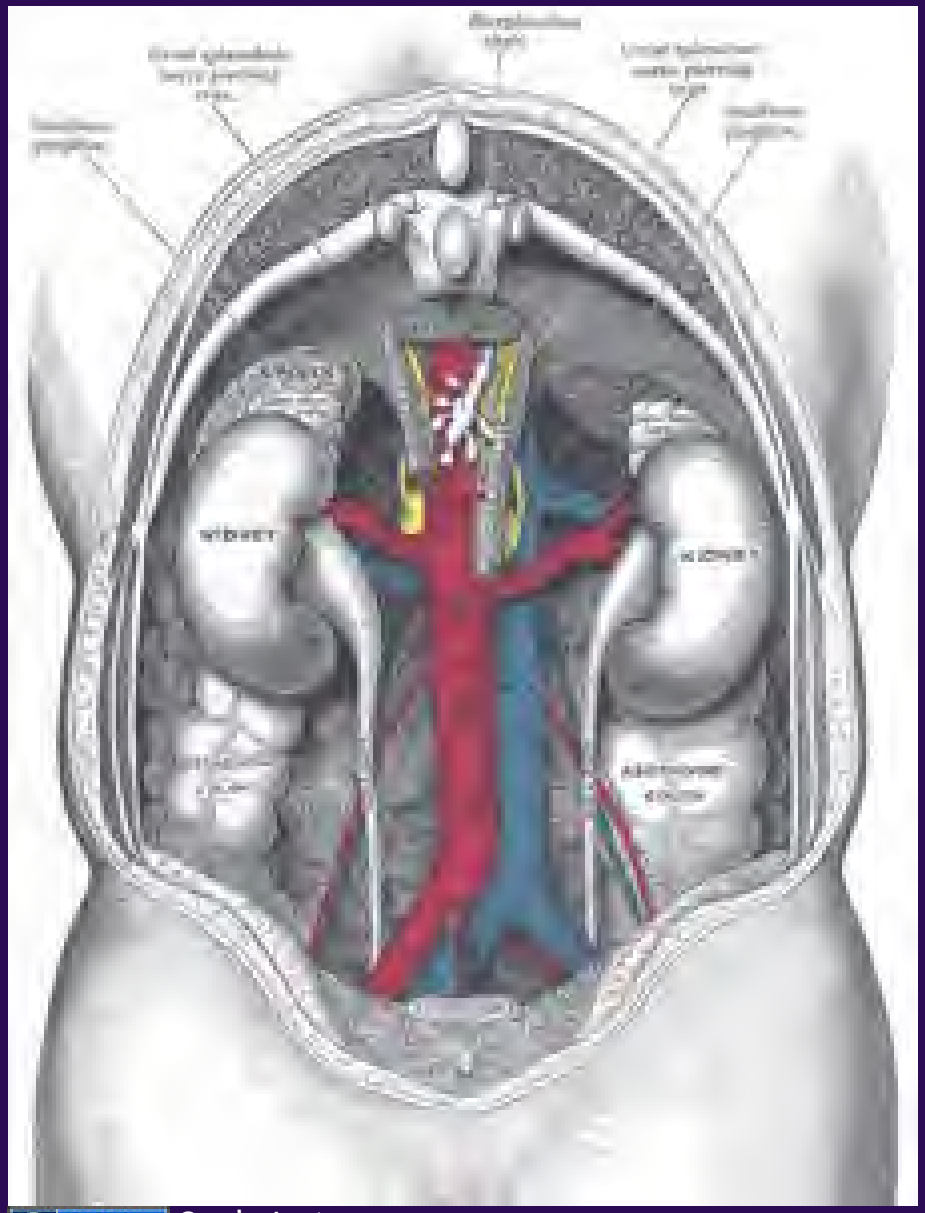


Anatomy of Kidney and Upper Urinary Tract

- "Tight spots" prone to obstruction
  - Ureteropelvic junction (UPJ)
  - Mid-ureter as crosses iliac vessels (over sacrum on plain radiograph)
  - Ureterovesical junction (UVJ)
- Of these, the UPJ most common site of congenital obstruction



PD-EXP Gray' s Anatomy



PD-EXP Gray’ s Anatomy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Case 1: Ureteropelvic Junction Obstruction (UPJO) Evaluation

- Intravenous urogram
  - Unilateral hydronephrosis and non-visualization of ureter
- Diuretic renal scintigraphy
  - 50% split renal function
  - T 1/2 (time for ½ of tracer to exit kidney) on symptomatic side > 100 minutes (normal < 10 minutes)

Intravenous Urogram

PD-INEL Source Undetermined

Left hydronephrosis and non-visualization of the ureter

Diuretic Renal Scintigraphy

Excretion from left kidney is delayed

Case 1: Ureteropelvic Junction Obstruction (UPJO) Treatment

- Percutaneous endopyelotomy
  - A minimally-invasive alternative to formal pyeloplasty
- Nephro-ureteral stent capped off in 1 week, and removed in 6 weeks
  - Complete resolution

Kidney & Upper Urinary Tract: Obstruction

Causes of UPJO

- Primary
  - Histological disorganization
    - excess longitudinal muscle fibers (loss of normal organization)
    - increase in collagen
    - attenuation of muscle bundles
  - Crossing vessels, high insertion, kinks, bands
- Secondary
  - Traumatic scar, iatrogenic scar, external compression (think cancer!)

Evaluation of UPJO

- Determine presence and degree of obstruction
  - Is there obstruction, or just dilation?
  - Complete or partial obstruction?
- Determine renal function
  - If kidney not working well, may be little value in repairing
- Determine cause of obstruction

Kidney & Upper Urinary Tract: Obstruction

Dilation of the Urinary Tract

- Non-obstructive
  - Ureteral reflux, prior obstruction, extra-renal pelvis, diuresis
  - Stagnation may lead to infection and calculi, but usually innocuous
- Obstructive
  - Increased resistance to urine flow that produces increased proximal pressure and subsequent loss of organ function
  - Occasionally difficult to distinguish from non-obstructive

Massive Vesico-ureteral Reflux on Cystogram

Evaluation of UPJO

- Anatomic tests
  - Intravenous urography
  - Renal (surface) ultrasonography
  - Computed tomography
  - Endoluminal ultrasonography
- Functional tests
  - Ultrasonography with resistive indices
  - Diuretic renal scintigraphy
  - Whitaker test

Intravenous Urography

Renal Ultrasonography

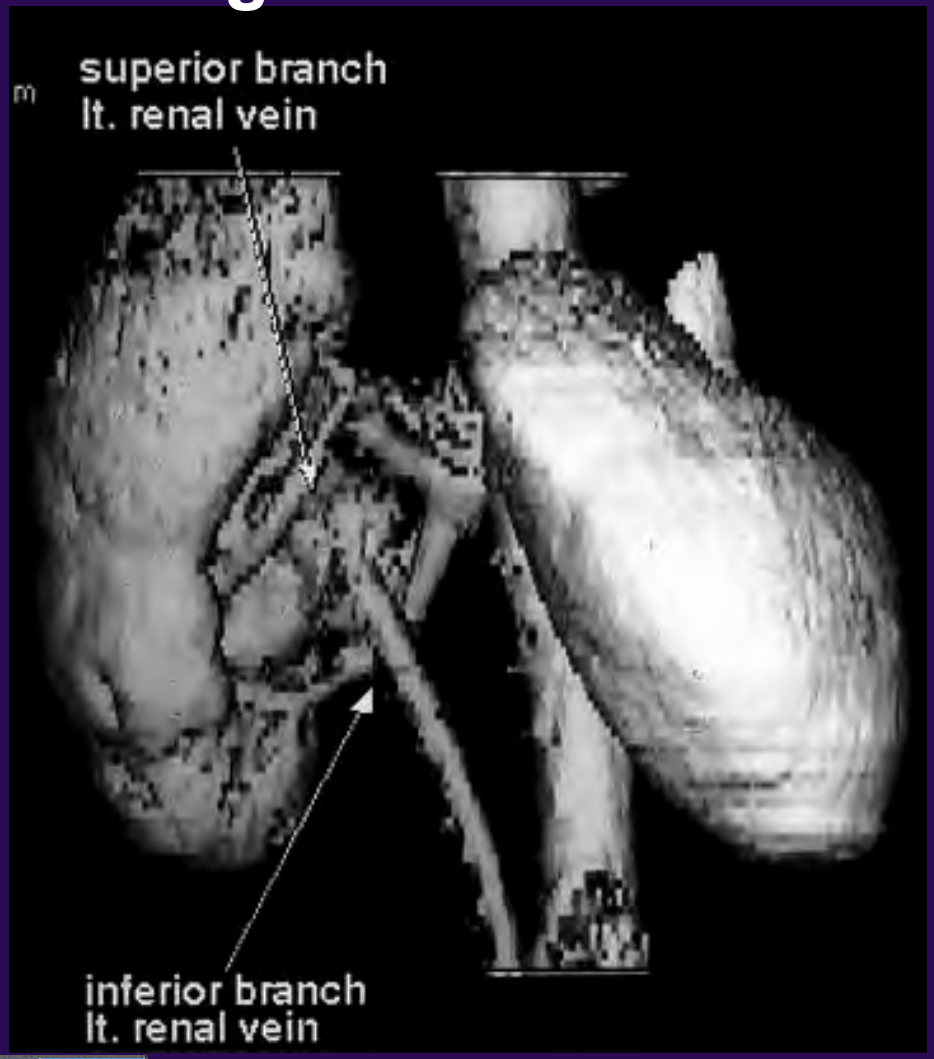
Computed Tomography

Can determine additional anatomy, including vessels crossing at UPJ

PD-INEL Source Undetermined

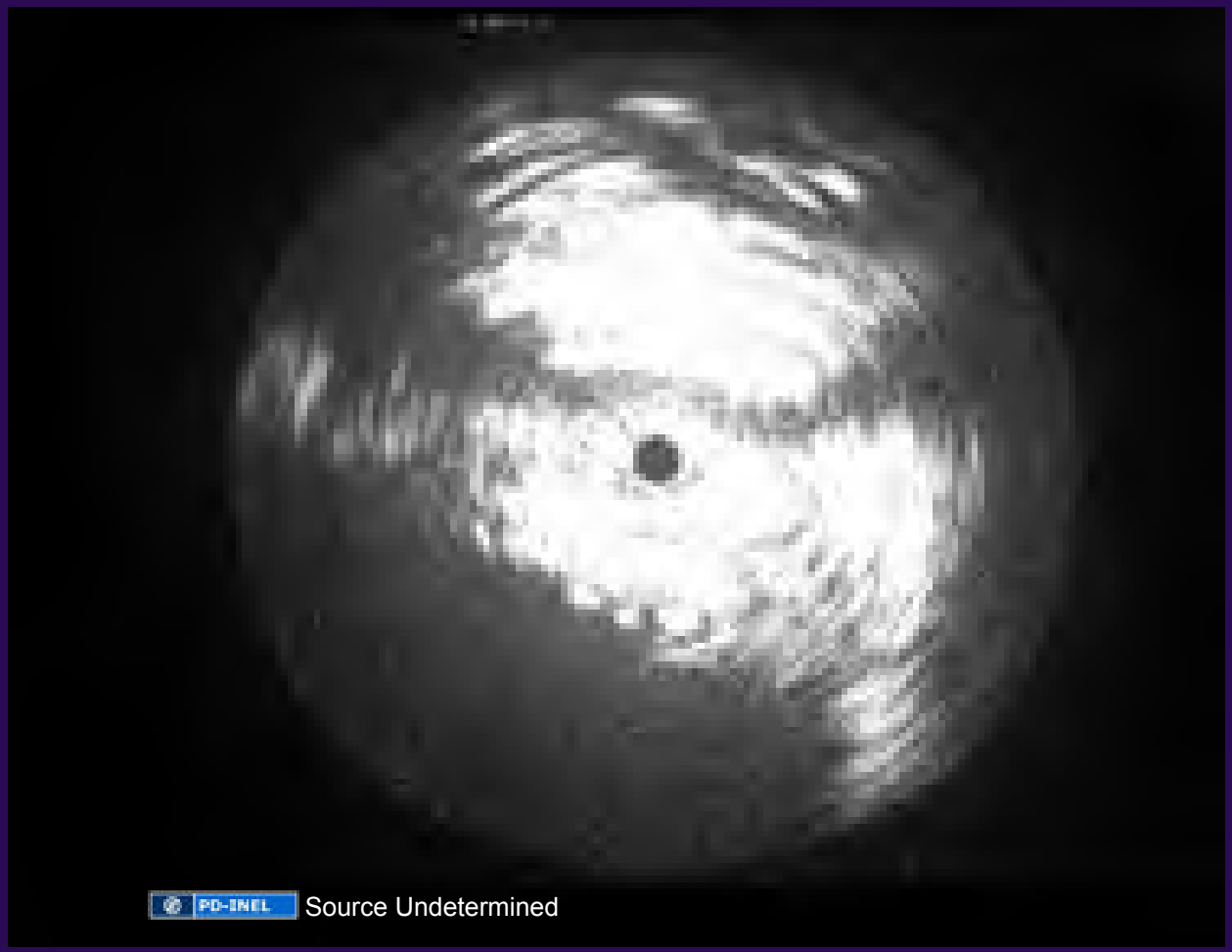
Computed Tomography

Can determine additional anatomy, including vessels crossing at UPJ

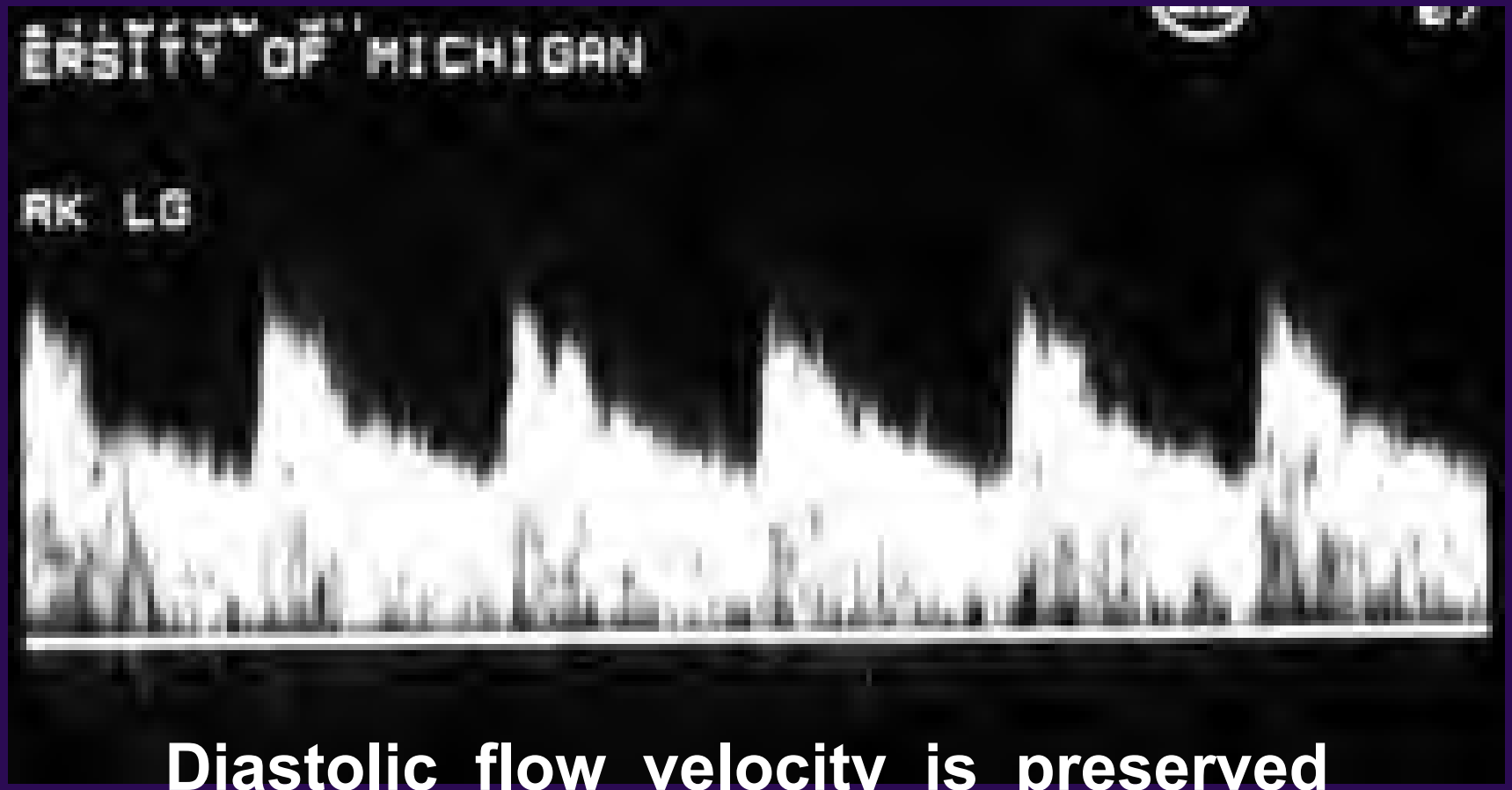


Endoluminal Ultrasonography

Can detect vessels crossing over UPJ, but requires retrograde catheterization

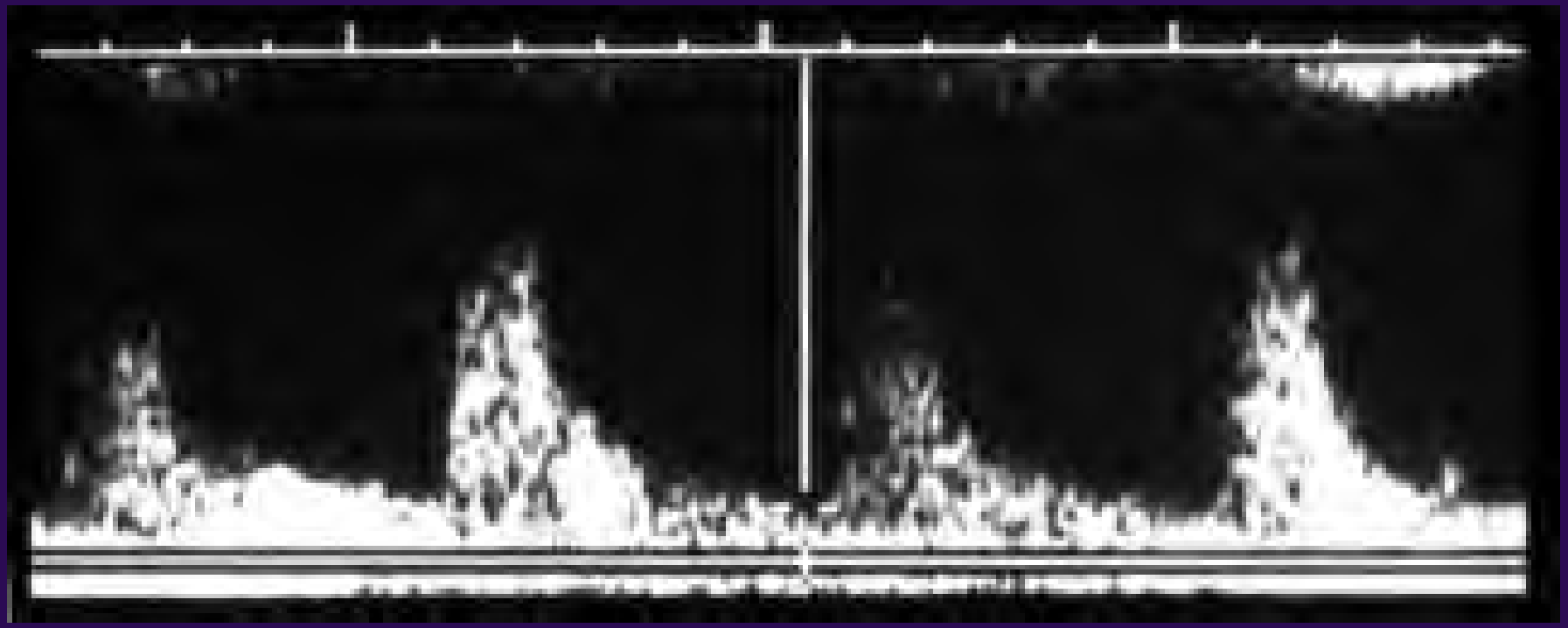


Ultrasonography with Resistive Indices (PSV - LDV) /PSV: Normal



Diastolic flow velocity is preserved

Ultrasonography with Resistive Indices (PSV - LDV) /PSV: Obstructed



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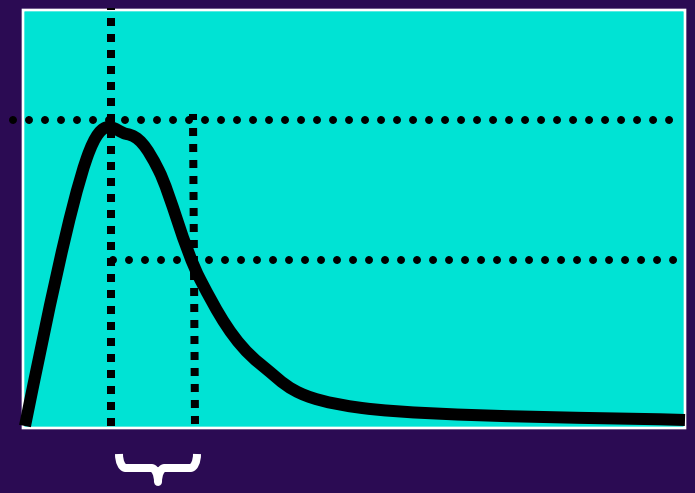
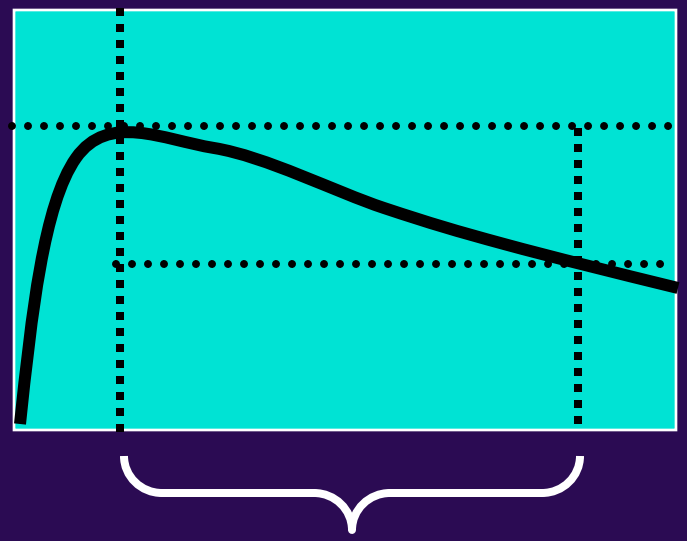
As resistance to blood flow increases, diastolic flow velocity decreases, and resistive index increases (R.I. > 0.70)

Diuretic Renal Scintigraphy: most definitive non-invasive test

T ½ = Time for ½ of radiotracer to be excreted from kidney after furosemide



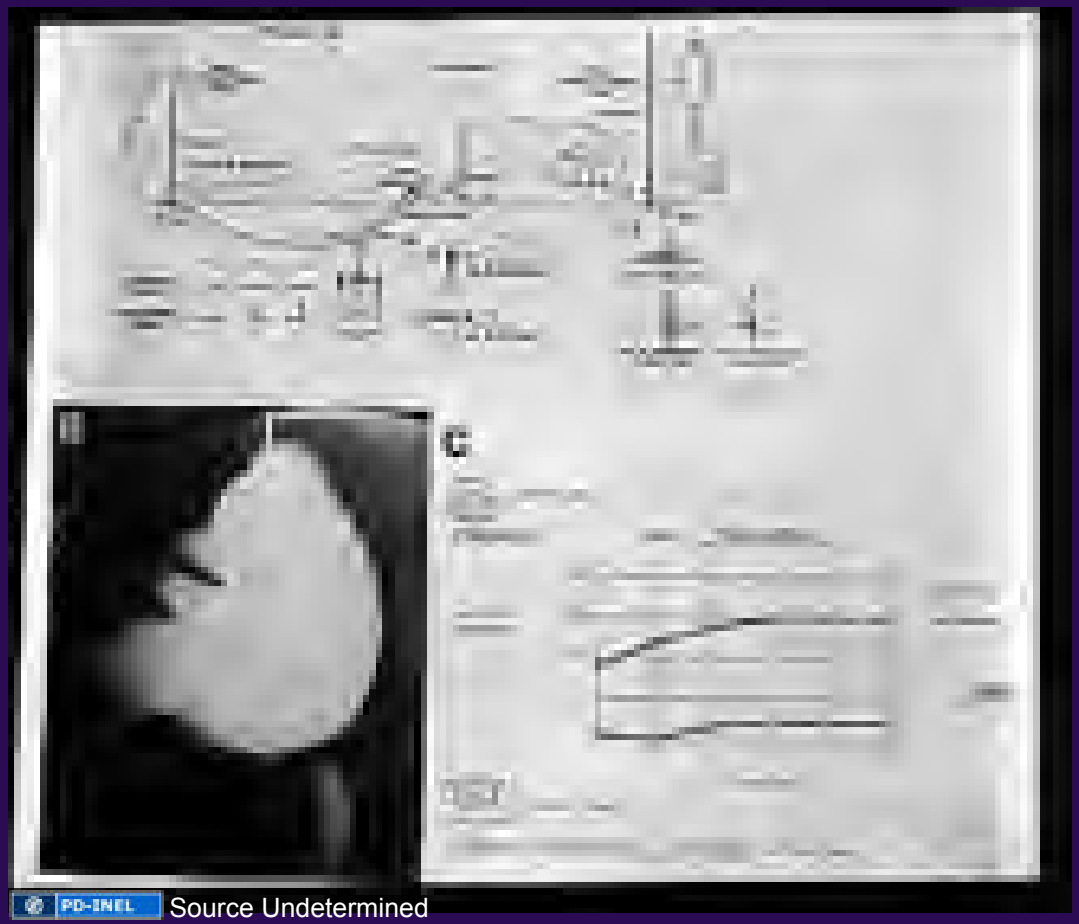
T ½ = 5 minutes
(normal < 10 min.)



T ½ = 25 minutes
(obstructed > 15 min.)

Whitaker Test: most definitive test

Pressure in renal pelvis at set infusion rate through nephrostomy tube (> 15 mmHg at 15 cc/min infusion = obstruction)



Kidney & Upper Urinary Tract: Obstruction

Treatment of UPJO

- Open pyeloplasty
- Laparoscopic pyeloplasty
- Percutaneous endopyelotomy
- Retrograde balloon dilation
- Retrograde endopyelotomy
- Acucise® (cutting balloon) endopyelotomy

Most Invasive / Most Effective

Least Invasive / Least Effective

Break

Case 2: Acutely Obstructing Distal Ureteral Calculus

- 30 year old man
- Sudden onset of right flank pain
- Initial gross hematuria, but now clear
- No fevers and chills, but nausea
- Frequent urination
- Pain somewhat less after a few hours
- Restless, moving about
- Right flank, lower quadrant, and testicular pain / tenderness
- 5 - 10 RBC / hpf on urinalysis

Case 2: Acutely Obstructing Distal Ureteral Calculus

- Sudden onset pain - c/w renal colic
- Urine cleared - suggests obstruction
- Nausea - very common with renal colic
- Freq. urination - irritation by distal calculi
- Pain decreased - forniceal rupture, decrease in pressure from renal hemodynamics
- Moving about - NOT peritonitis
- Flank to scrotum - expected radiation
- Urinalysis - RBCs in 85%

Kidney & Upper Urinary Tract: Calculi

Suspected Acute Ureteral Obstruction Differential Diagnosis

- Intrinsic
  - Calculi, tumor, clot, edema
- Extrinsic
  - Compression by tumor, lymph node
- Acute versus acute-on-chronic
  - Calculi, or calculi impacted into partially-obstructing stricture?
  - Clot, or clot on tumor?

Suspected Acute Ureteral Obstruction Radiographic Evaluation

- KUB and Intravenous urography
  - Anatomic picture, localize pathology
- Ultrasonography
- CT
  - Non-contrast (stones)
  - CT urogram (with contrast, like IVU)
- Retrograde pyelography
  - Injection through catheter

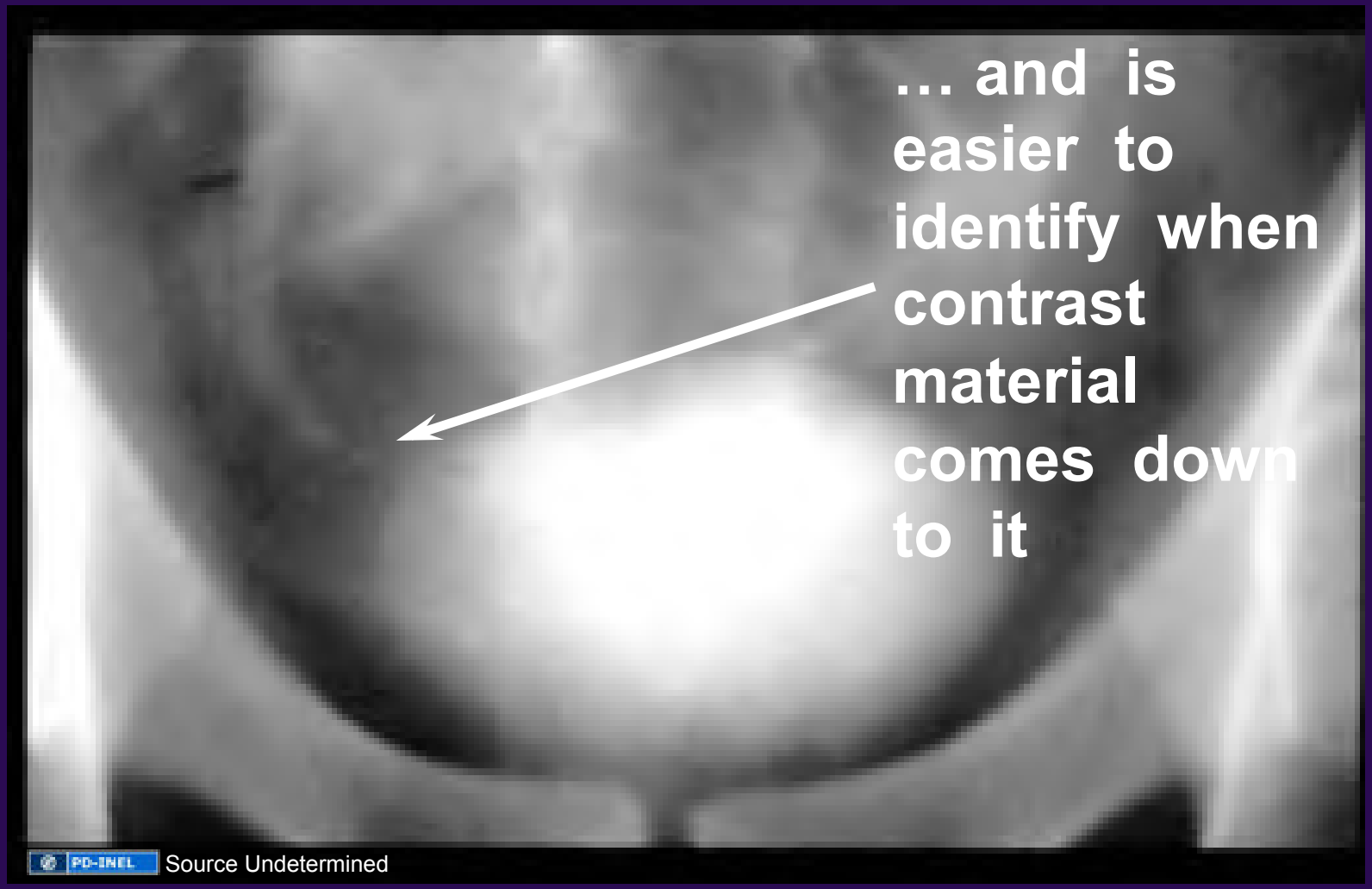
Intravenous Urography

PD-INEL Source Undetermined

This stone is less dense than contrast material, so appears as filling defect

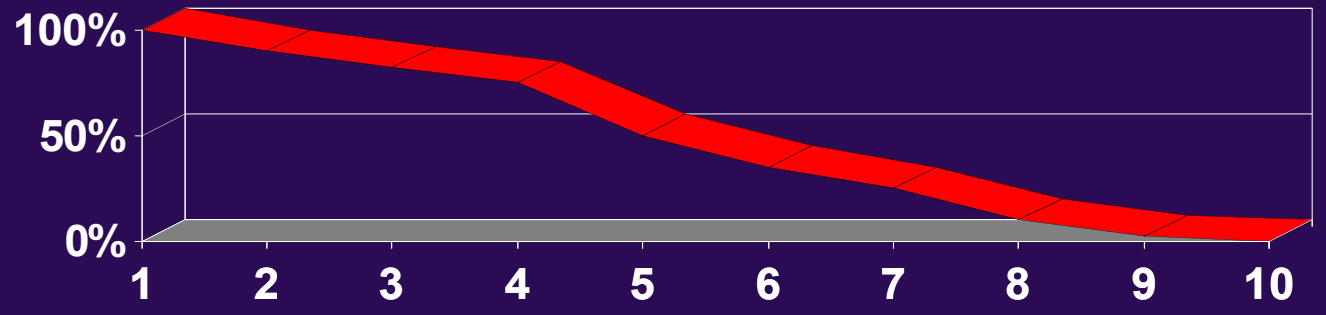
Intravenous Urography

Intravenous Urography

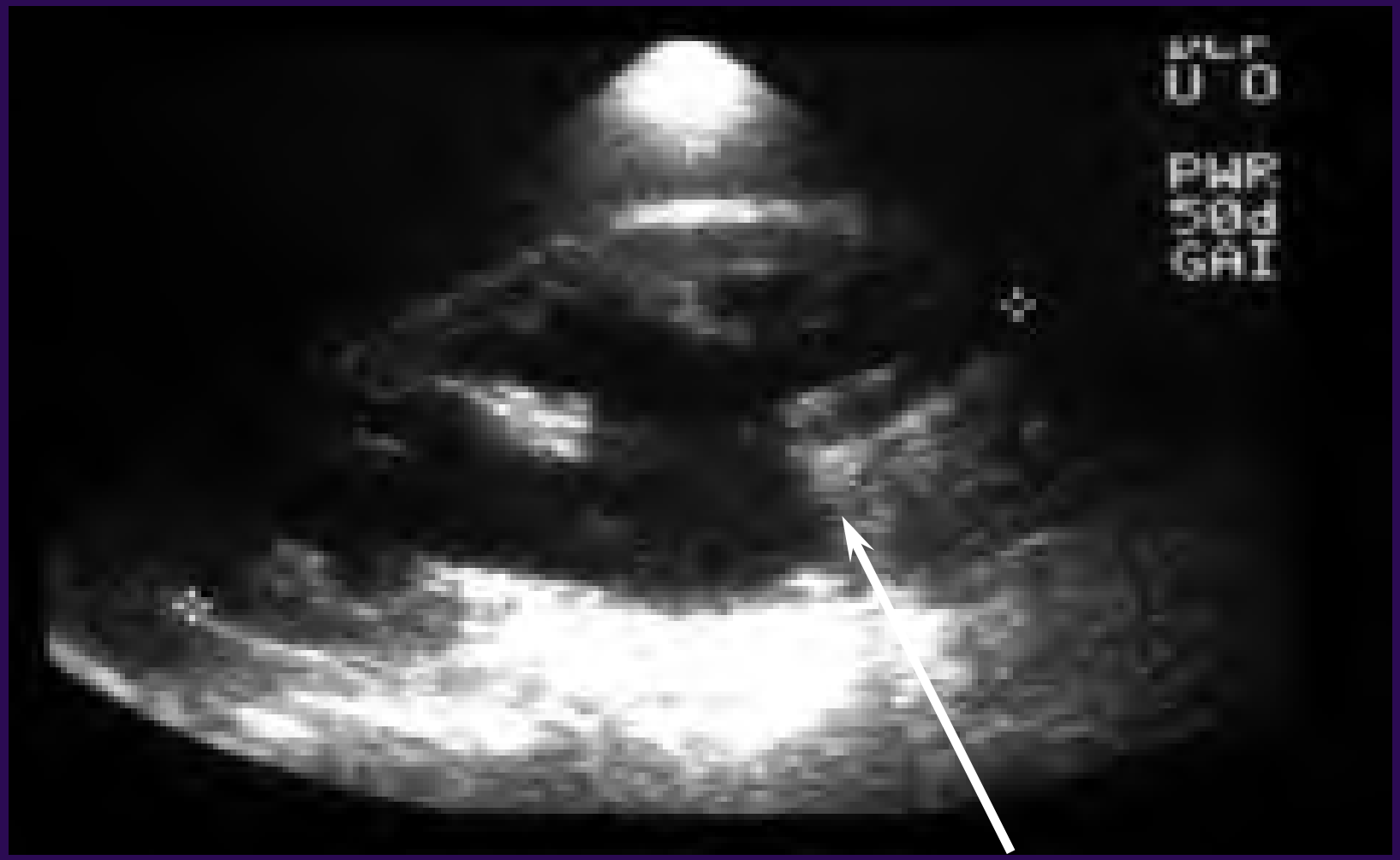


Spontaneous Passage of Ureteral Stones

Width	Proximal	Middle	Distal
4 mm	20%	45%	55%
5 mm	6%	30%	45%
6 mm	0%	10%	25%



Ultrasonography

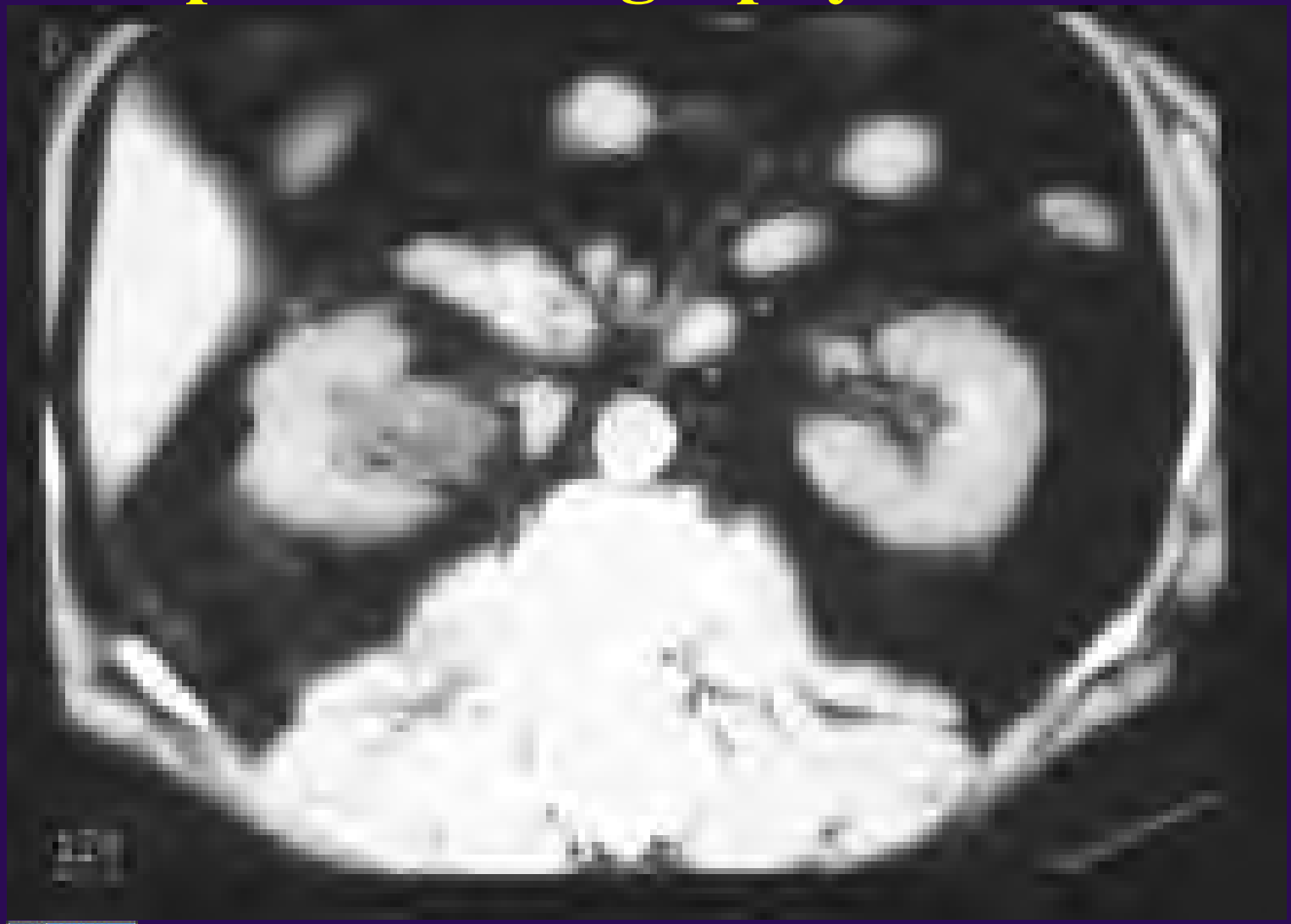


Hydronephrosis

Computed Tomography

(almost) all stones dense on CT

Computed Tomography



Secondary signs of ureteral obstruction

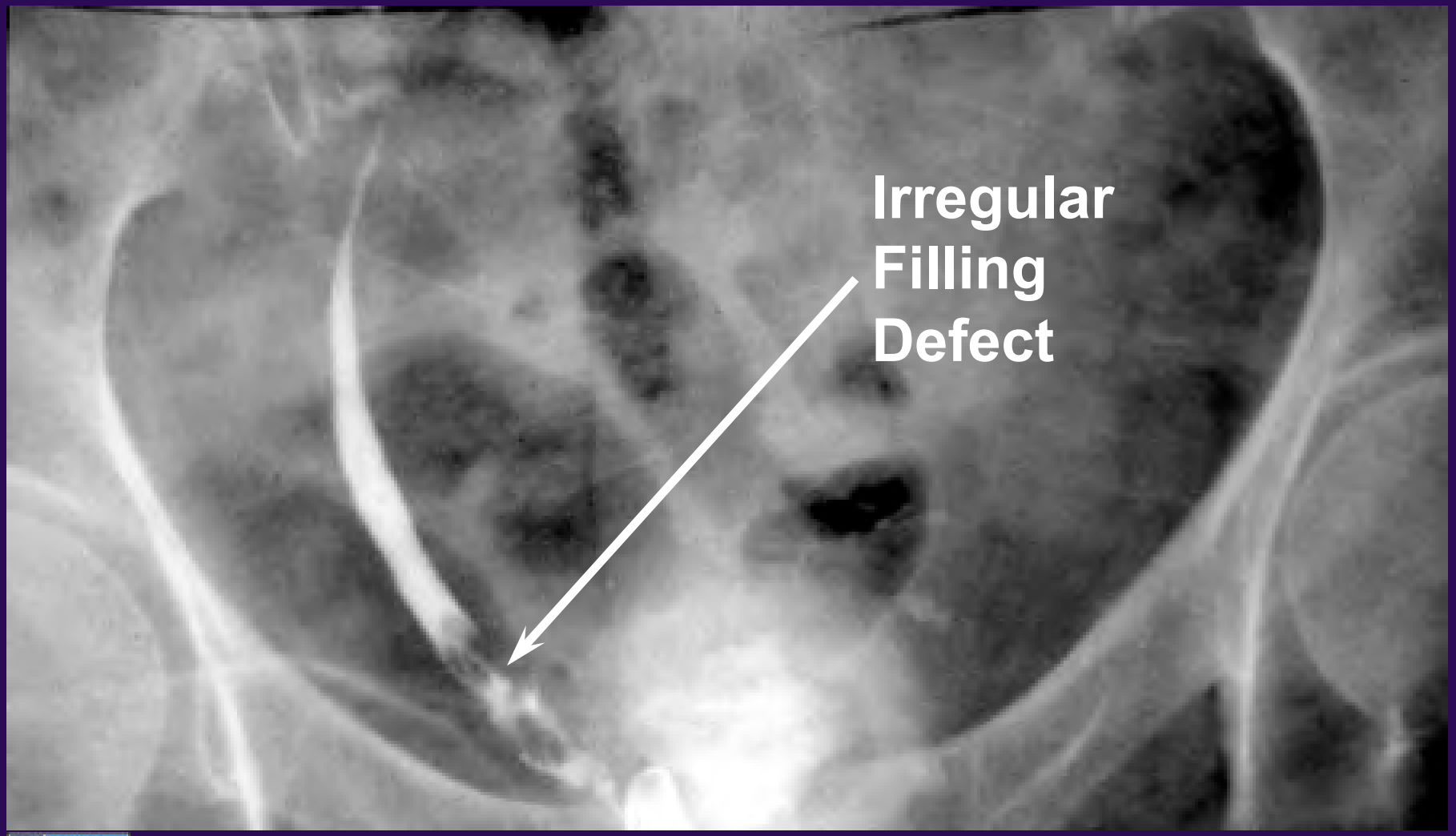
Computed Tomography

Secondary signs of ureteral obstruction

Computed Urography

PD-INEL Source Undetermined

Intravenous Urography



Kidney & Upper Urinary Tract: Calculi

If "filling defect" on contrast study …

- Neoplasm
  - Urothelial neoplasm most common
- Blood clot
  - Will resolve during follow-up
- "Radio-lucent" calculus (15%)
  - Other 85% are calcium containing
  - Refers to appearance on plain film (all typical stones are opaque on CT scan)
  - Radio-lucent stones are usually uric acid (only medically dissolvable stone)
- Need to rule-out tumor!

Case 3: Non-obstructing Renal Calculus

- 65 year old man
- Microscopic hematuria
- Remote history of urolithiasis
- Mild prostatism
- Unremarkable PE except for prostatic enlargement
- Urinalysis - 10 RBC / hpf

Plain Radiography

Densely radio-opaque stones

Indications for Surgical Treatment of Urolithiasis

- Urinary tract infection
- Significant obstruction
- Pain refractory to oral medication
- Others
  - Staghorn calculi - risk of urosepsis
  - Long-standing ureteral calculi - eventual obstruction
  - Occupational or lifestyle reasons

Plain Radiography

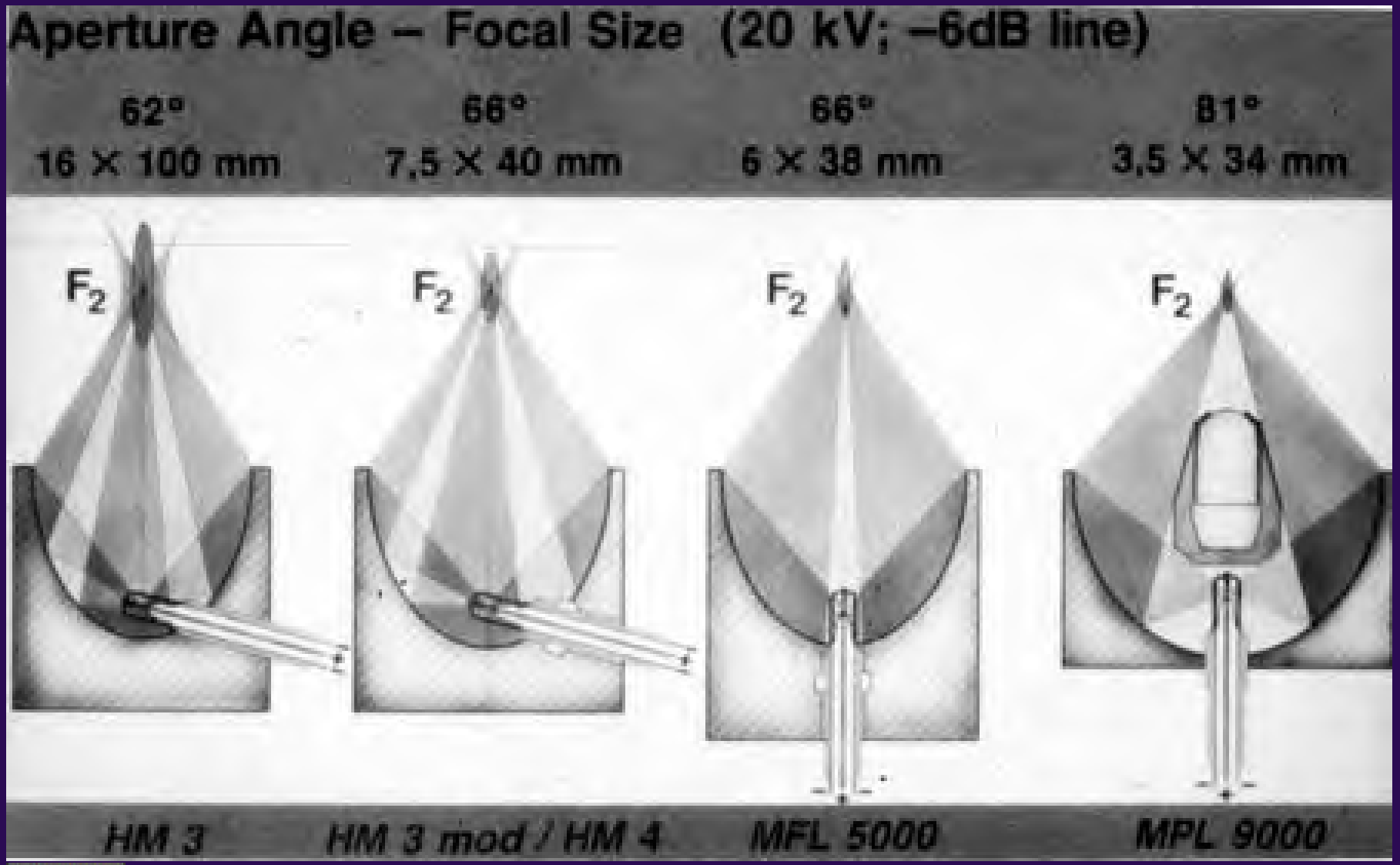
Staghorn Calculus

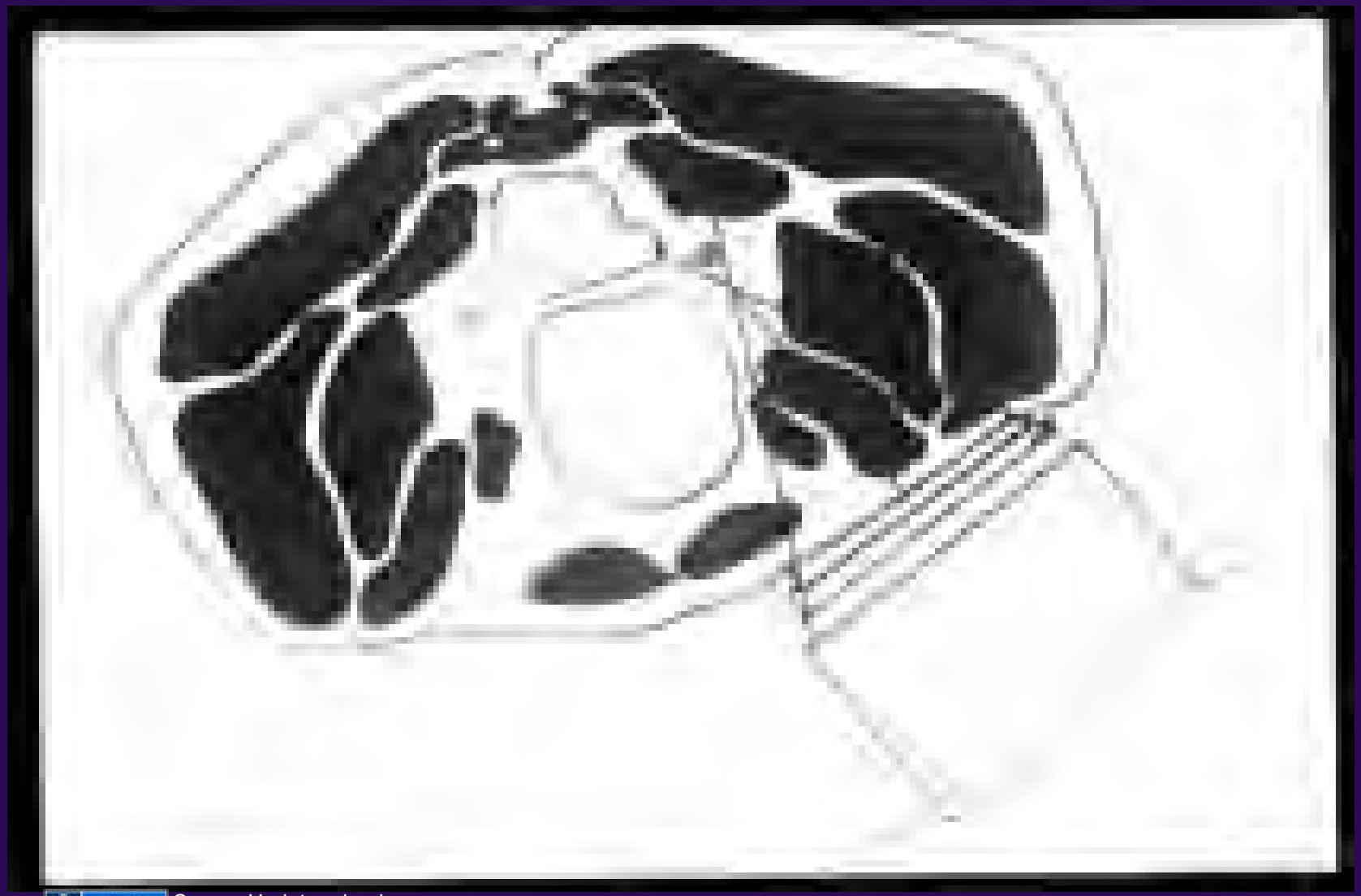
Surgical Treatment of Urolithiasis

- Open surgical / laparoscopic lithotomy
- Percutaneous nephrostolithotomy (Antegrade endoscopy)
- Ureteroscopy (Retrograde endoscopy)
- Extracorporeal shock wave lithotripsy (SWL)

Most Invasive / Most Effective

Least Invasive / Least Effective



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Parameters Determining Treatment of Urolithiasis

- Size
- Location
- Composition
- Medical Condition, Patient Preference, Physician Preference

Kidney & Upper Urinary Tract: Calculi

Size

- Small
  - SWL
- Moderate
  - Ureteroscopy
- Large
  - Percutaneous nephrostolithotomy

Kidney & Upper Urinary Tract: Calculi

Location

- Distal Ureter
  - Scope > SWL
- Middle Ureter
  - Scope > SWL
- Proximal Ureter
  - SWL > Scope ?
- Kidney
  - SWL > Scope

Computed Tomography

Distal aspect of UVJ, almost in bladder

Computed Tomography

Huge bilateral renal calculi

Kidney & Upper Urinary Tract: Calculi

Composition

- Dense (Calcium oxalate monohydrate)
  - Scope
- Fuzzy/ Faint (Calcium oxalate dihydrate, calcium phosphate, struvite)
  - SWL
- Cystine
  - Scope
- Uric Acid
  - SWL

Plain Radiography

Densely radio-opaque stone

Plain Radiography

Case 4: Pyelonephritis

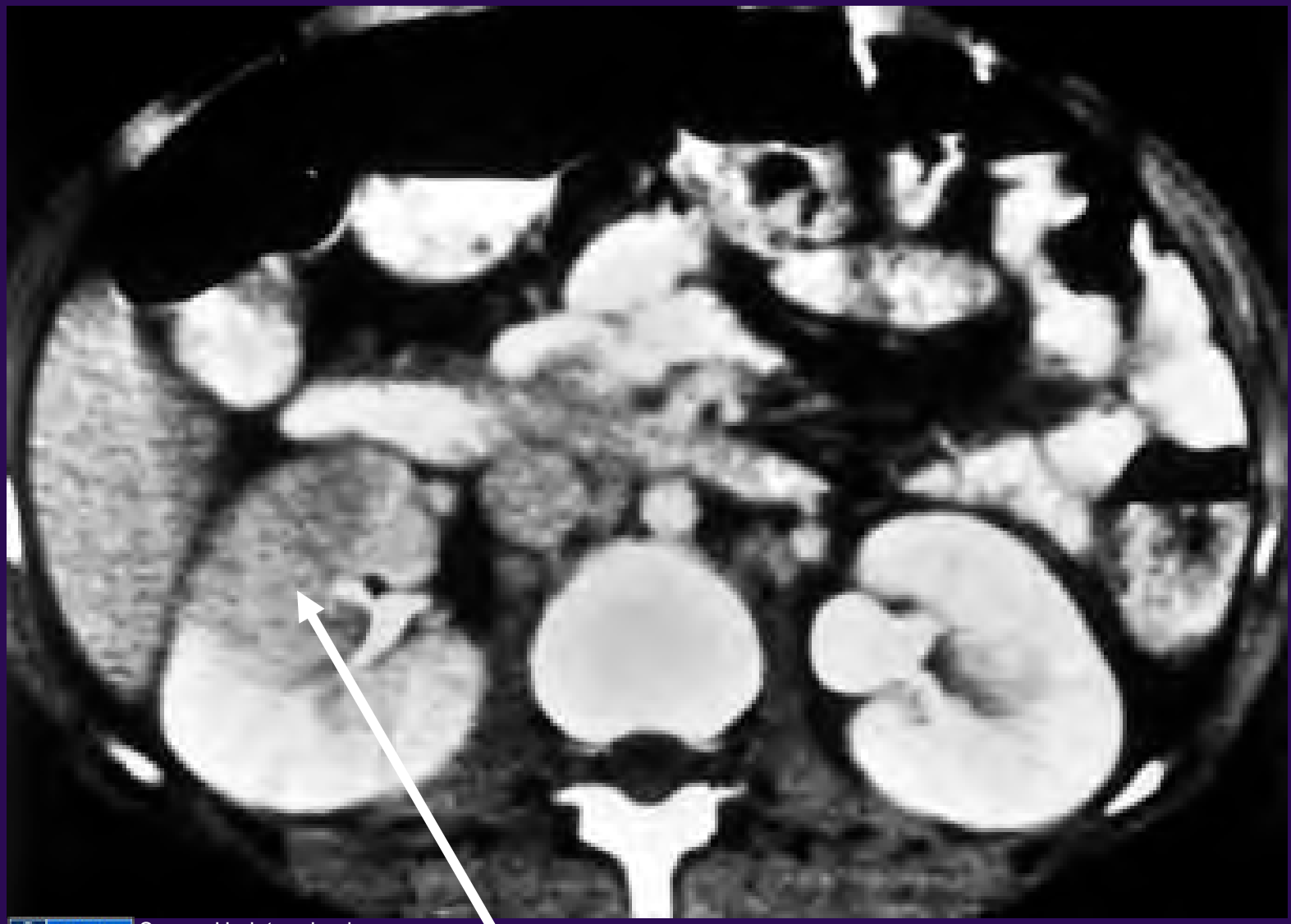
- 29 year old woman
- Started with 3 days of dysuria (painful urination), urinary frequency / urgency
- Now temp 101.7º C, right flank pain
- History
  - Occ previous UTI
  - Recently married
  - Limited sexual activity before marriage, now active
- PE - Right CVAT

Case 4: Pyelonephritis

- Young woman - UTI is common
- Initial cystitis (infection ascends) common and suggest UTI
- Systemic symptoms - distinguishes upper from lower tract UTI
- Previous UTI - helps establish her characteristic symptoms of UTI
- Sexual activity - predisposing factor for UTI in women
- CVAT - renal involvement

Acute Pyelonephritis

- Most common disease of the kidney
- Usually ascending infection
- Diagnosis by clinical findings and urine culture (85% are GNR; E. Coli)
- Imaging used to detect complications or to assess for predisposing factors
- Complications: papillary necrosis, pyonephrosis, abscess, sepsis
- Predisposing factors: obstruction, calculi, vesico-ureteral reflux



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Lobar Nephronia

Management of Acute Pyelonephritis

- Typical case - fever resolves within 48 hrs of starting antibiotics
  - Oral abx in most
  - Intravenous abx if very ill-appearing
- Debilitated patients at greater risk
  - Diabetes mellitus
  - Steroids
  - Chronically ill
  - Immuno-suppressed

Management of Acute Pyelonephritis

- History of complications or lack of response to antibiotics
  - → US or CT for obstruction, calculi
- Upper tract obstruction with infection
  - → DRAINAGE (stent, percutaneous tube)
- Upper tract calculus with infection
  - → Treat acute infection, then calculus
- If child or recurrent
  - → US for scars
  - → Voiding cystogram for reflux

Kidney & Upper Urinary Tract: Pyelonephritis

Drainage procedures

- Percutaneous nephrostomy tube

Image of a percutaneous nephrostomy tube removed

- Ureteral stent

Break

Case 5: Renal Cell Carcinoma (RCC)

- 64 year old man
- Gross hematuria
- Flank pain
- History
  - 66 pack-year smoker
- PE
  - Fullness in right flank

Case 5: Renal Cell Carcinoma (RCC)

- Older man - peak in 6th decade, 3:1 M:F overall
- Hematuria - most common presenting sign
- Flank pain and flank mass - along with hematuria, the classic triad
- Smoking - major acquired risk factor

Demographics of RCC

- Incidence: 12.8 per 100,000 (‘00 - ‘04)
- Mortality: 4.2 per 100,000 (‘00 - ‘04)
- estimated 51,190 new cases in 2007 (~2% of adult malignancies)
- estimated 12,890 deaths in 2007
- Lifetime Risk (M / F)
  - Risk of occurrence - 1.71 / 1.01 %
  - Risk of death - 0.59 / 0.35 %

SEER Cancer Statistics

Kidney & Upper Urinary Tract: Masses & Cysts

Risk factors for RCC

- Cigarettes
- Obesity
- Hypertension
- Occupational exposures
- Dialysis
- Hereditary
  - von Hippel-Lindau disease
  - Tuberous Sclerosis

Pathology of RCC

- Proximal tubular cell neoplasm
- Often venous involvement
- Hemorrhagic, necrotic, cystic, and calcified components common
- Metastasize most commonly to lung, liver, bone, adrenal, and contralateral kidney

Kidney & Upper Urinary Tract: Masses & Cysts

Common Renal Tumors

Pathology	Malignant?	Relative %
Renal Cell Ca.	Yes	85%
Urothelial Ca.	Yes	5%
Oncocytoma	No	5%
Other	Most not	5%

Histology

- Most Renal Cell Carcinomas have "Clear cell" histology
  - Lipids (dissolve out during slide processing) and glycogen
  - Fuhrman grading (1 to 4)
    - 1 = well differentiated
    - 4 = poorly differentiated

Genetics

- Most renal cell carcinomas are sporadic
- Are associated with several syndromes, the most common of which is Von-Hippel Lindau syndrome
  - Autosomal dominant
  - Cerebellar and retinal vascular tumors
  - Adrenal and renal tumors (inc cysts)

Genetics

- Von-Hippel Lindau syndrome
  - Autosomal dominant
  - Mutation in VHL tumor suppressor gene: 3p25-26
- 95% of sporadic “clear cell” renal cell carcinomas have VHL mutation
  - One of the strongest associations among solid tumors
  - Opportunities for gene therapy

Symptoms / Signs of RCC

- Hematuria (29 – 60%)
- Flank pain (14 – 51%)
- Flank mass (21 – 47%)
- All 3 = Classic triad
  - Present in < 10%
  - Usually signifies advanced disease

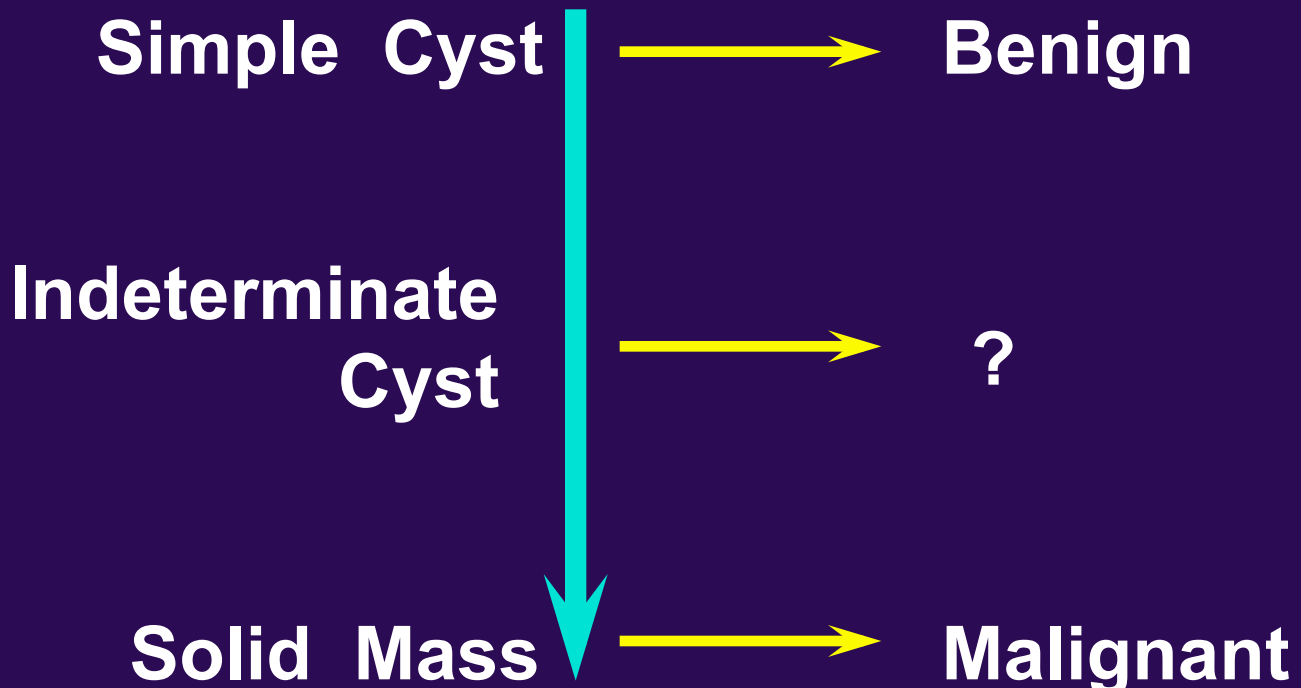
Kidney & Upper Urinary Tract: Masses & Cysts

Paraneoplastic Syndromes

- ESR elevation
- Calcium elevation
- Polycythemia
- Anemia
- Thrombophlebitis
- Hyperinsulinism
- LFT elevation

Kidney & Upper Urinary Tract: Masses & Cysts

Central Role of Imaging



Stage Migration of RCC due to more frequent imaging

- 1970's
  - 4% of RCC ≤ 3 cm
  - 32% presented with metastases
- 1980's
  - 25 - 40% were incidental finding
  - 25% of RCC ≤ 3 cm
  - 17% presented with metastases
- 1990's
  - 60% were incidental finding

Kidney & Upper Urinary Tract: Masses & Cysts

Imaging Modalities

- Intravenous Urography (IVU, IVP)
- Ultrasonography (US)
- Computed Tomography (CT)
- Magnetic Resonance Imaging (MRI)

Intravenous Urography

- Typical upper tract imaging for hematuria work-up
- Excellent visualization of collecting system
- Renal mass or cyst causes displacement of surrounding organ or deformation of outline
- Screening study only
- If renal mass / cyst suspected, further imaging is required

Intravenous Urography

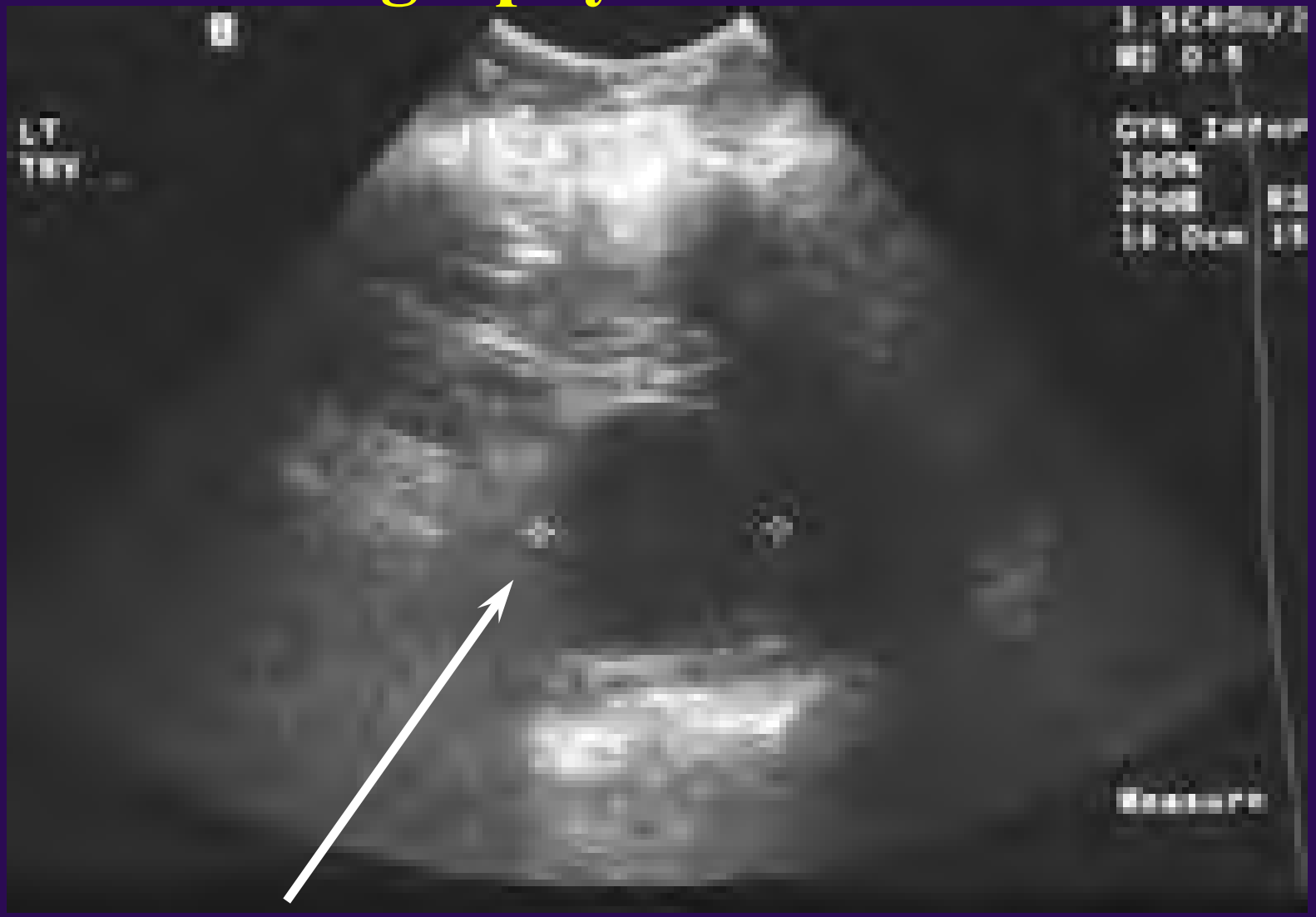
Mass Effect in kidney

Kidney & Upper Urinary Tract: Masses & Cysts

Ultrasonography

- Usual follow-up to IVU suspicious for renal mass / cyst
- No ionizing radiation
- Non-invasive
- Operator dependent
- Reliably identifies simple cyst (85% of renal mass / cysts)
- If NOT a simple cyst
  - ➡ Cross-sectional imaging

Ultrasonography

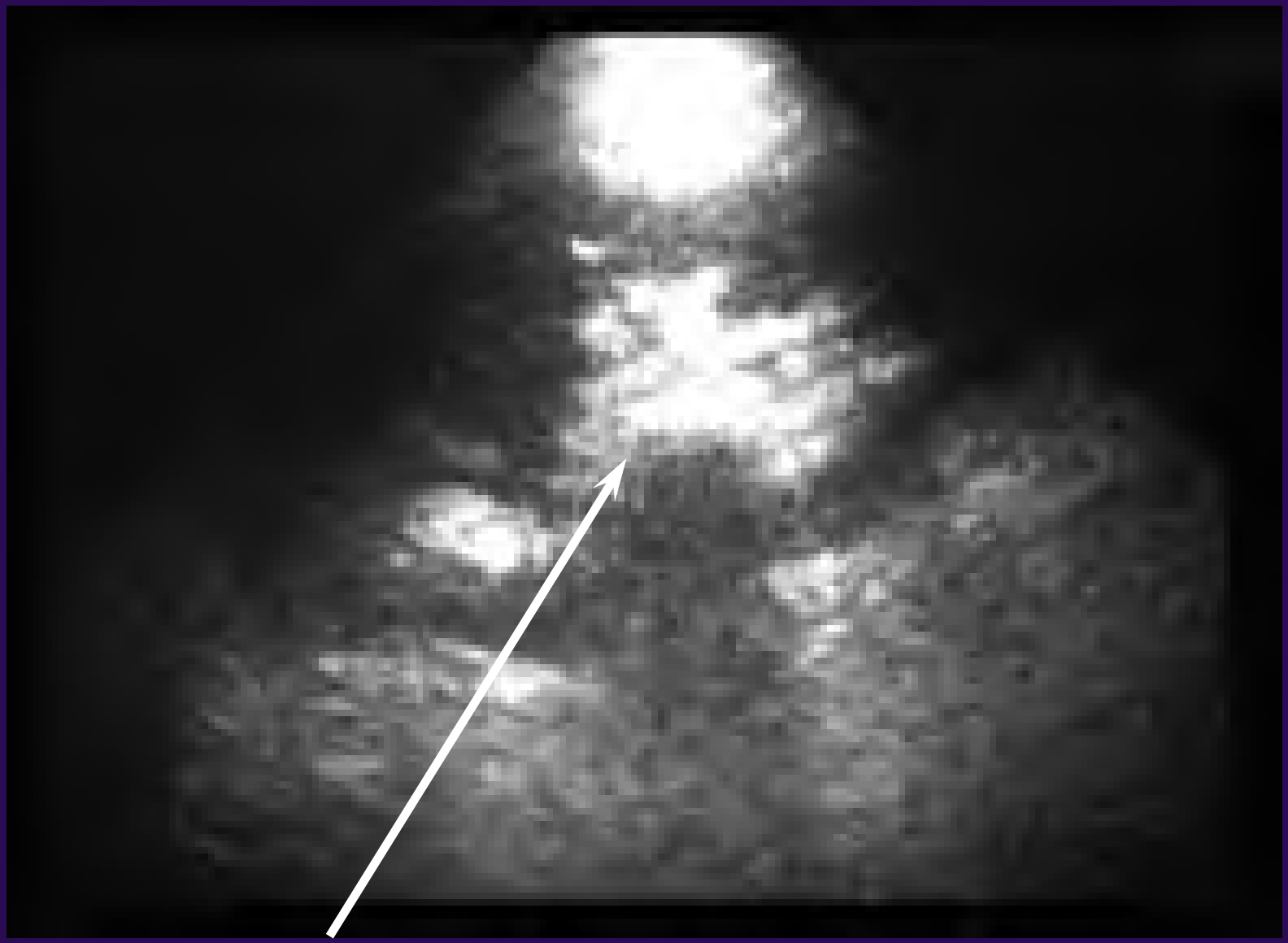


Simple Cyst

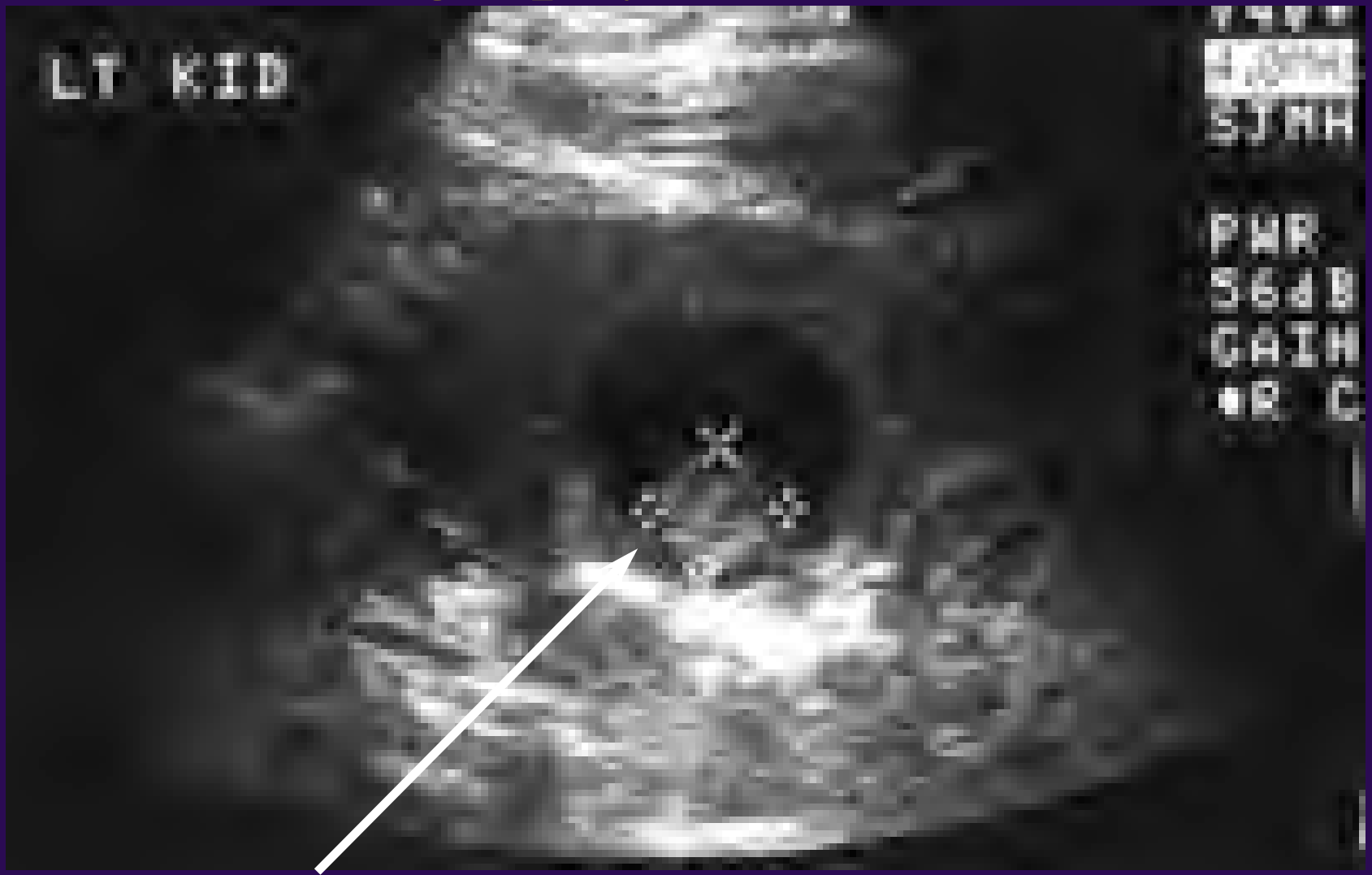
Sonographic Characteristics

- Anechoic with enhanced through transmission
  - ➡ Simple cyst
- Echogenic with acoustic shadowing
  - ➡ Stone or other calcification
- Iso / hypoechoic mass
  - ➡ Solid mass

Ultrasonography

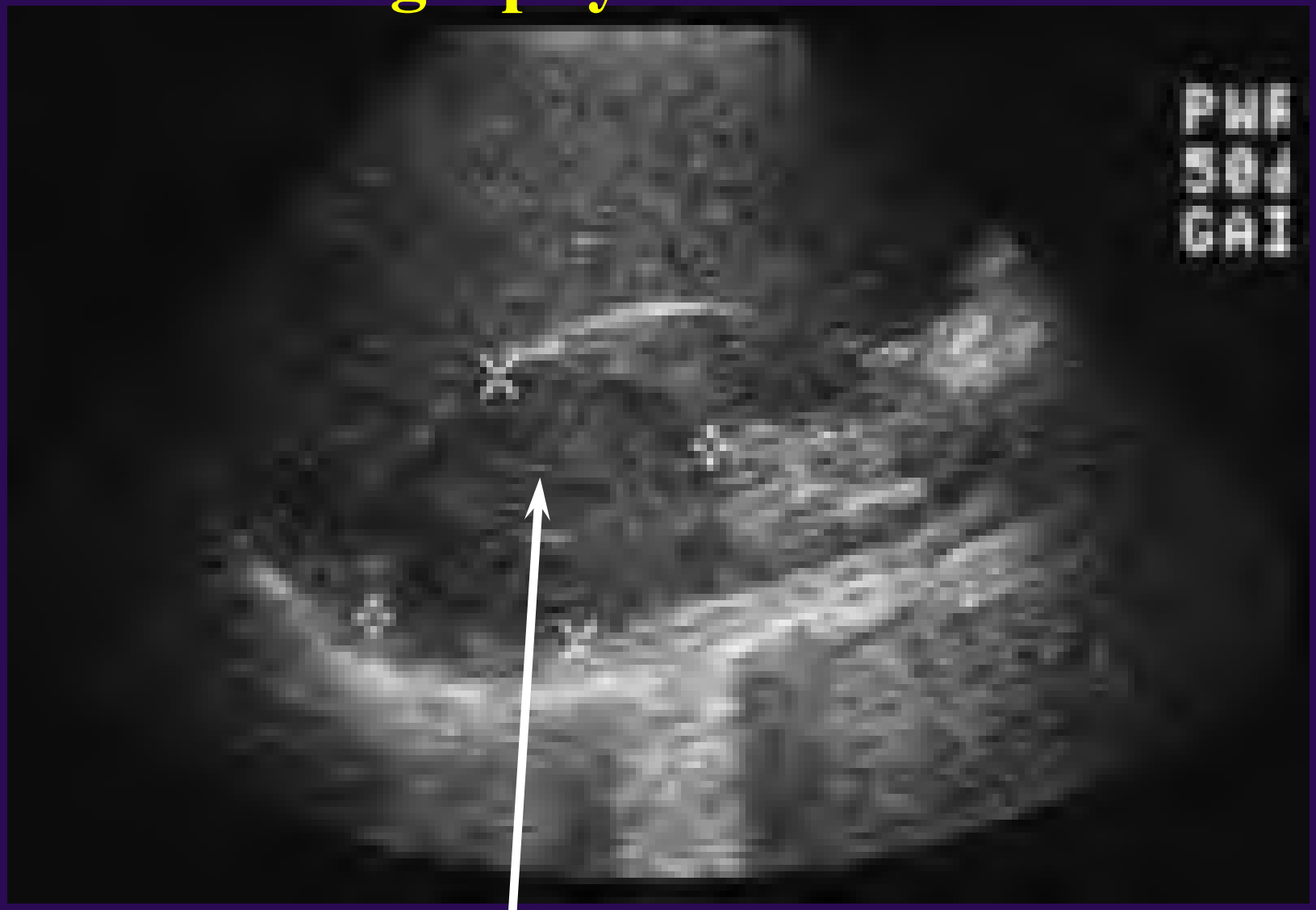


Ultrasonography



Nodule in Cyst

Ultrasonography



Solid Mass

Computed Tomography

- Current gold standard
- Non-contrast scan, then scan with intravenous contrast
  - Enhancement = Hounsfield units (density) increase by ≥ 10 with contrast
- 3 to 5 mm maximum cut width
- Spiral CT - single breath hold
  - Minimize motion artifact
  - Exact duplication of cuts

Computed Tomography

PD-INCL Source Undetermined

Simple Cyst

Computed Tomography

Complex Cyst

Renal Cell Carcinoma

Kidney & Upper Urinary Tract: Masses & Cysts

Solid, Enhancing Renal Mass on CT is RCC until Proven Otherwise

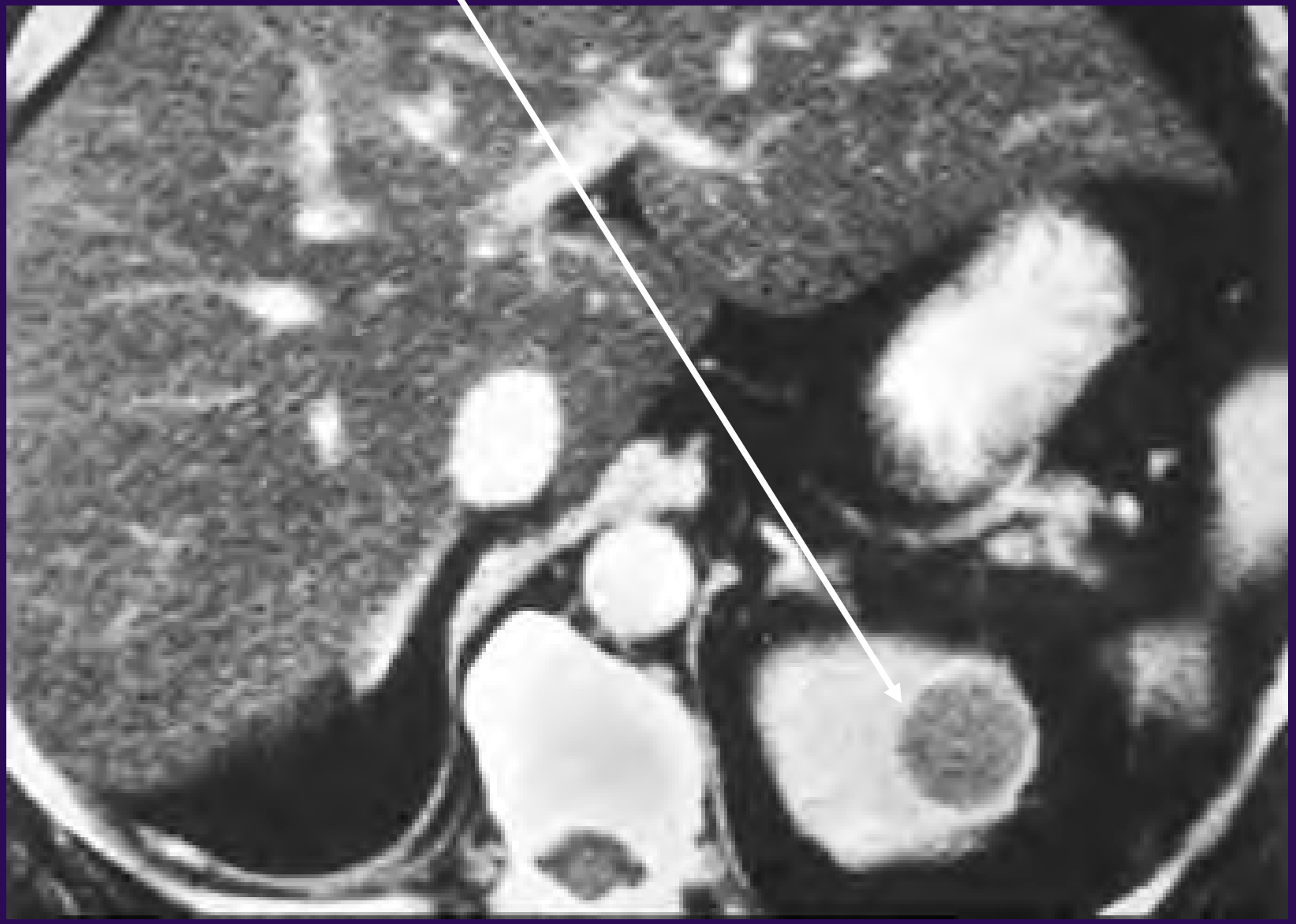
- Other possibilities
  - Oncocytoma
    - Benign, but indistinguishable from RCC on imaging
  - Angiomyolipoma
    - Benign, but can bleed if large
    - Usually diagnosed by imaging fat
  - Inflammatory mass
    - History of febrile illness
  - Lymphoma
    - Malignant, but no surgery

Oncocytoma ?

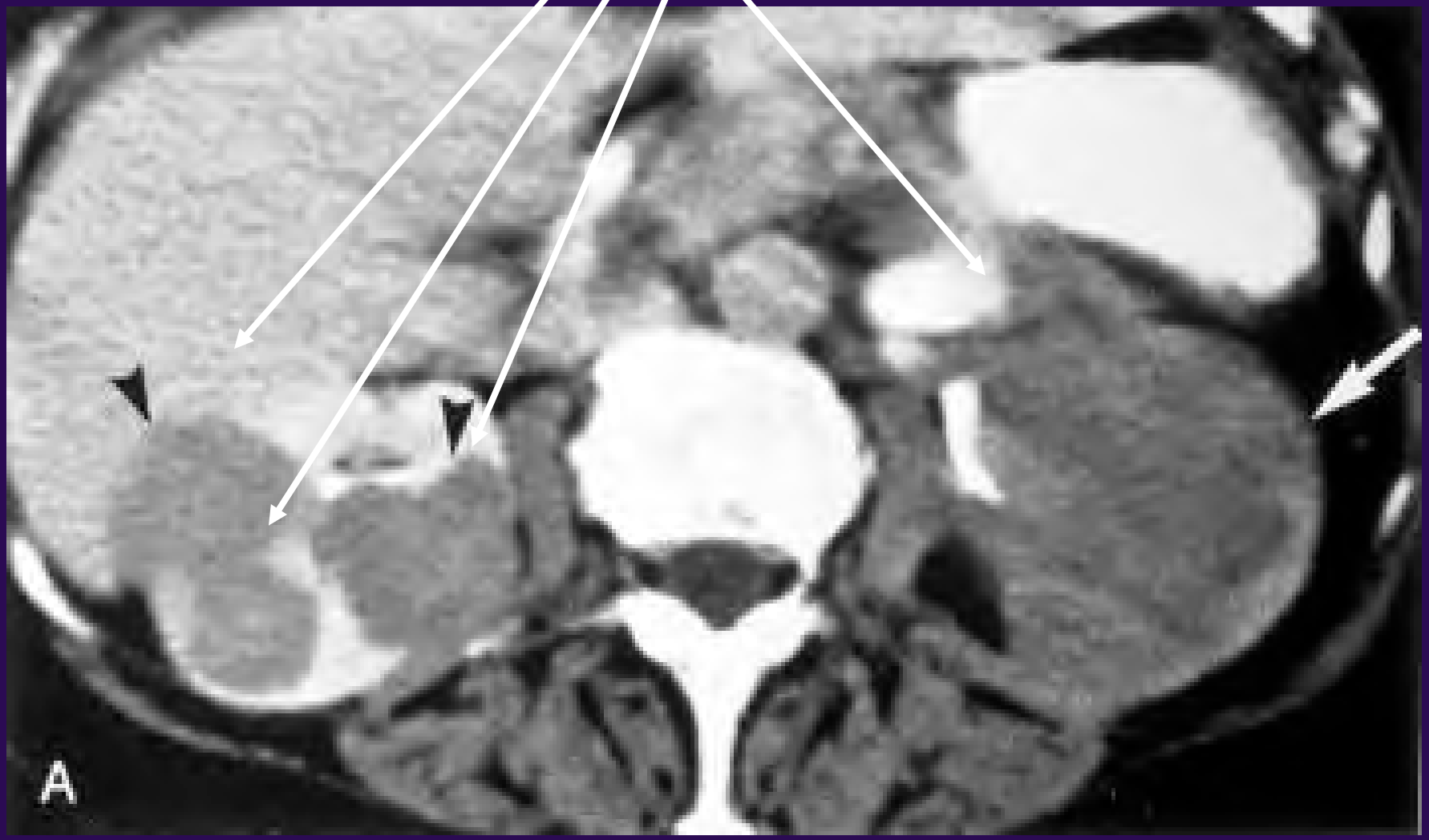
PD-INEL Source Undetermined

Angiomyolipoma

Renal Abscess



Renal Lymphoma



Magnetic Resonance Imaging

- Currently no advantage over CT except in certain situations
  - Allergy to contrast material
  - Elevated creatinine
  - Distinguish wall in some cysts
  - Detection of venous tumor thrombus in RCC (has replaced invasive venography)

Magnetic Resonance Imaging

Magnetic Resonance Imaging

Tumor Thrombus in Vena Cava

Sensitivity for Diagnosis of RCC $\leq$ 3 cm

- Intravenous urography - 67%
- Ultrasonography - 79%
- Computed tomography - 94%

Evaluation of Suspected RCC

- Initial imaging study (often IVU or US, either incidentally or for workup of hematuria or other signs / symptoms)
- CT or MRI for local assessment
  - Define lesion
  - Assess nodes, vein, other organs
- Staging
  - CXR, Bloods, + Bone Scan and others

Staging and Management of RCC

- Stage I
  - Tumor < 7 cm limited to kidney (T1)
  - no LN+ or metastases
  - ➔ Radical or Partial Nephrectomy
- Stage II
  - Tumor > 7 cm limited to kidney (T2)
  - No LN+ or metastases
  - ➔ Radical Nephrectomy

Staging and Management of RCC

- Stage III
  - Tumor into vein, fat, or adrenal
  - Or one single LN+
  - ➔ Radical Nephrectomy
- Stage IV
  - Tumor beyond Gerota's fascia (T4)
  - Or > 1 LN+ Or metastases
  - ➔ Systemic Therapy (Chemo, Immuno)

Prognosis of RCC

	5 year survival
• Stage I	~90%
• Stage II	~80%
• Stage III	~40 - 60%
• Stage IV	~10%

Case 6: Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- 37 year old woman
- No recent medical care
- Left flank "fullness"
- Adopted, unknown family history
- PE
  - Mass in left flank
  - BP 170/100

Case 6: Autosomal Dominant Polycystic Kidney Disease (ADPKD)

- Middle-aged - usually diagnosed in third decade
- "Fullness" - HUGE bilateral cysts, symptomatic in ~15%
- Over 50% have family history
- Hypertension - almost always, given enough time

Kidney & Upper Urinary Tract: Masses & Cysts

ADPKD

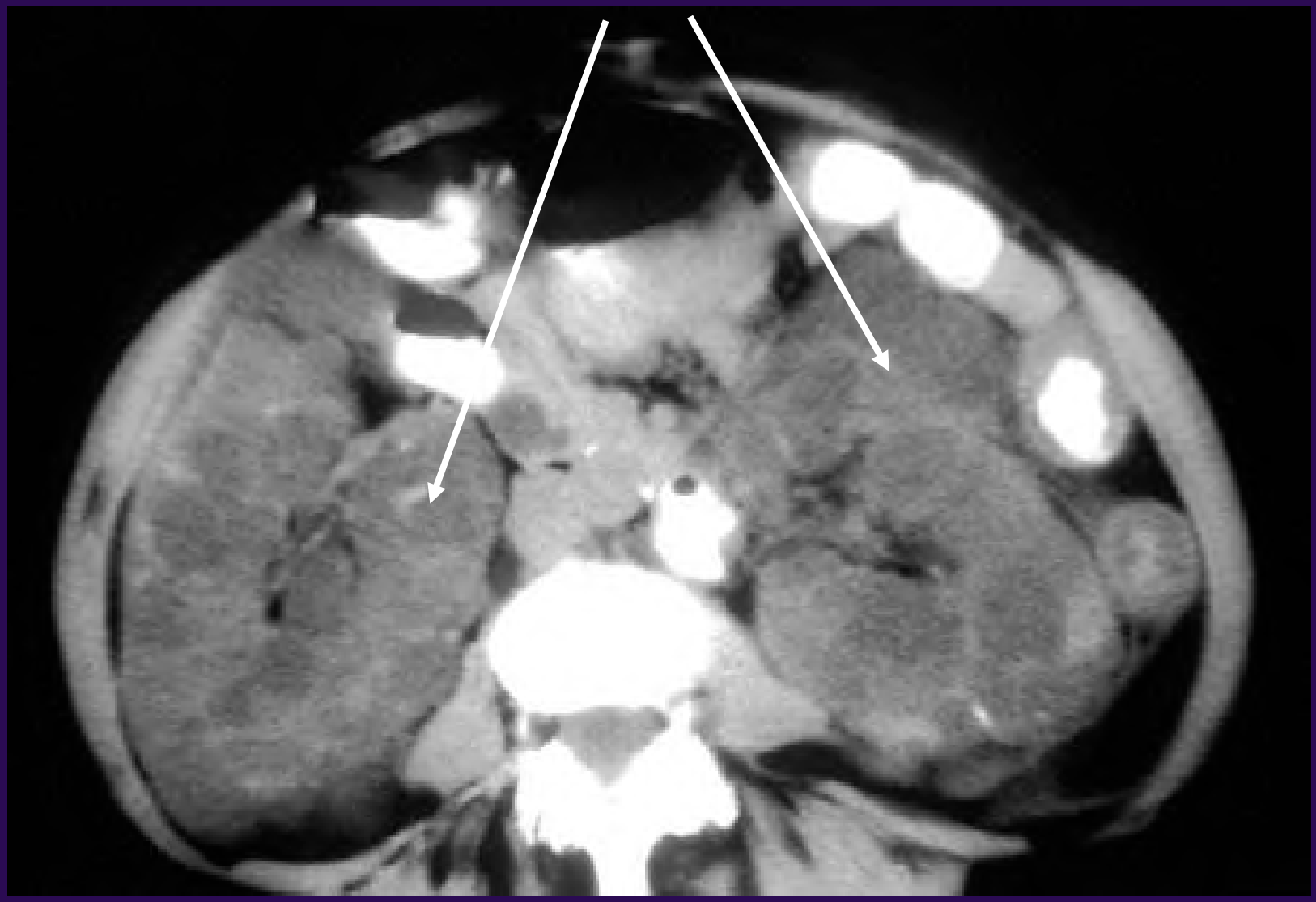
- Autosomal dominant inheritance
  - 85% from *PKD1* mutation (16q13)
  - 15% from *PKD2* mutation (4q21-23)
- Virtually 100% penetrance
- Incidence
  - 1 / 1000 live births
  - 6000 new cases annually
- Prevalence ~ 200,000

Kidney & Upper Urinary Tract: Masses & Cysts

ADPKD

- Epithelial proliferation in renal tube → renal tubule become cyst → as cyst grows it compresses renal parenchyma
- < 1% tubules become cysts
- Also hepatic, pancreatic and splenic cysts, and cerebral aneurysms
- Hypertension
- Renal failure - most but not all, usually in 5^{th} decade

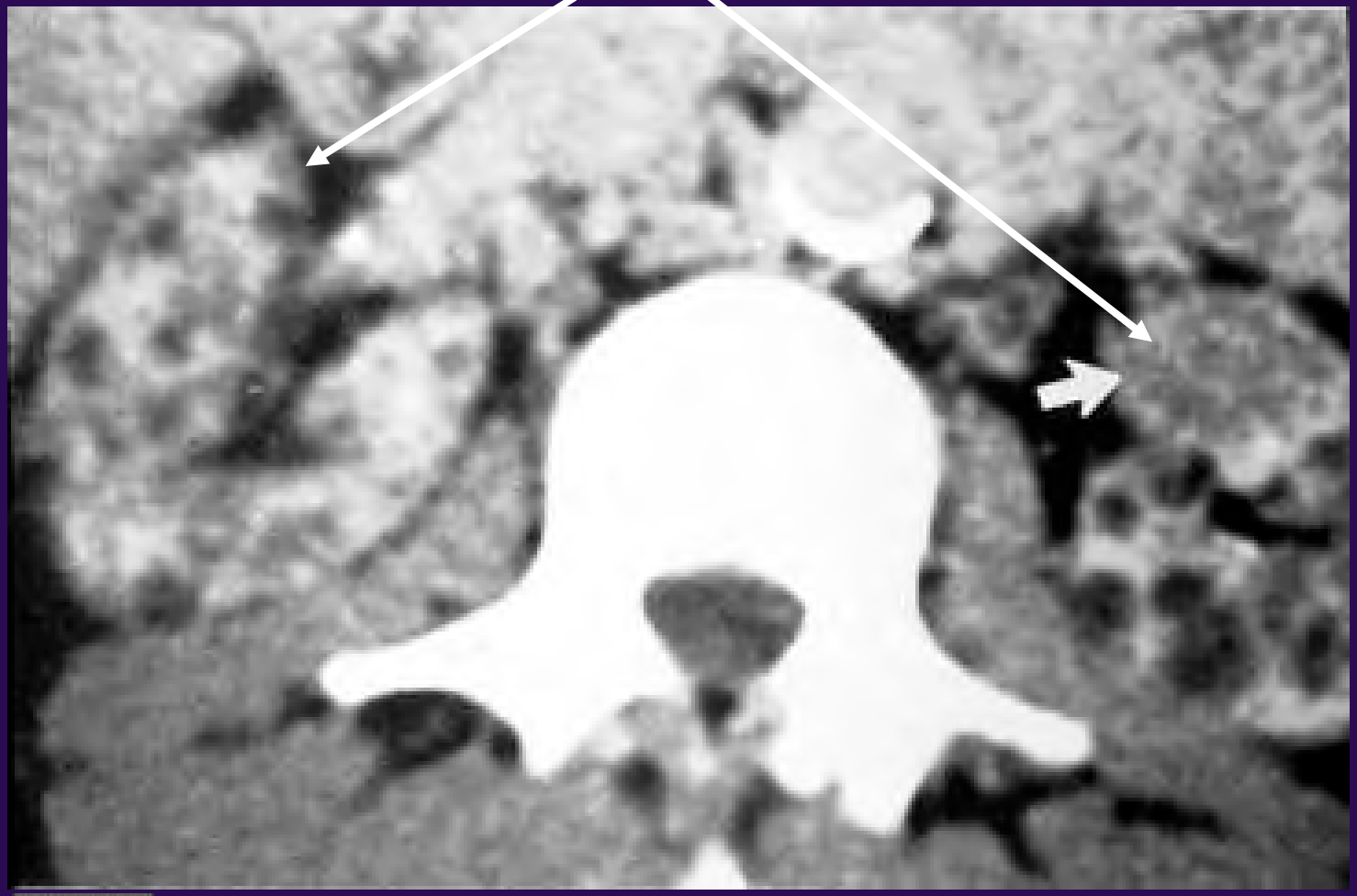
ADPKD



Other Cystic Diseases of the Kidney

- Simple cysts
  - In population over age 50
    - 50% pathologically, 33% by CT
  - Single or multiple
- Simple cyst complicated by hemorrhage or infection ("Complex cyst")
- Acquired cystic kidney disease (ACKD)
  - 40% of dialysis patients by 3 years
  - Controversial increase in RCC

ACKD



Evaluation of Renal Cysts

- Simple cysts definable with US
  - No further work-up needed
- ADPKD, ACKD
  - Definable by history and PE
- "Complex Cyst"
  - Simple cyst complicated by infection or hemorrhage
  - Cystic renal cancer
  - Diagnostic dilemma, surgery often required

Additional Source Information

for more information see: http://open.umich.edu/wiki/CitationPolicy

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Slide 30: Gray's Anatomy
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