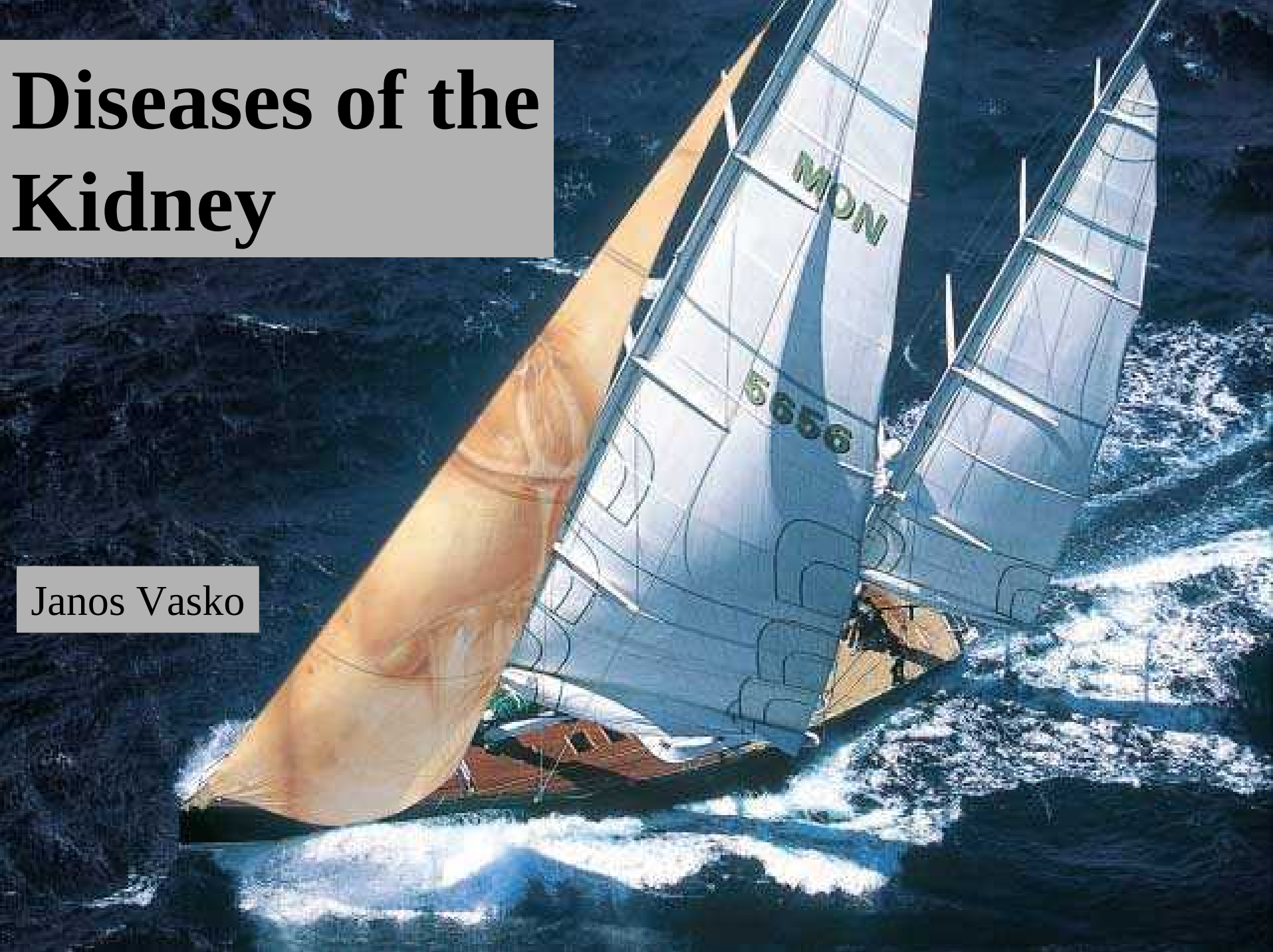
Diseases of the Kidney



Janos Vasko

- Congenital anomalies
- Glomerular diseases
- Tubulointerstitial diseases
- Infections
- Vascular diseases
- Stones
- Tumours

POLYCYSTIC KIDNEY DISEASE

- **INFANTILE TYPE**

- **ADULT TYPE**

  Autosomal dominant

  Cysts of varying size in both kidneys

  Normal parenchyma between cysts

  Cysts in liver, pancreas, spleen

  Intracranial aneurysmas

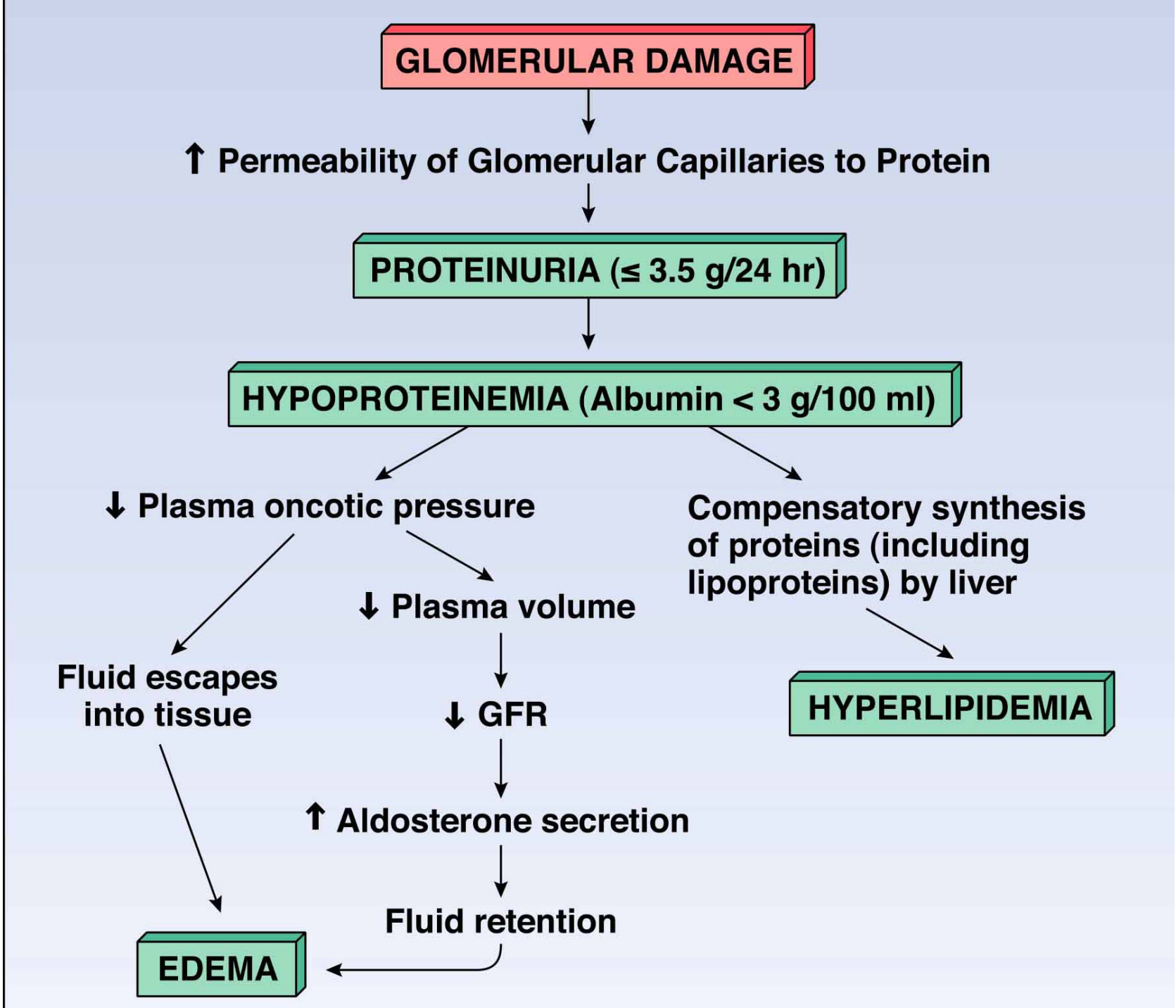
SIMPLE KIDNEY CYSTS

- Common findings at autopsy
- 50% at 50 years
- No clinical significance (except cancer diagnosis)

GLOMERULAR DISEASES

- **Nephrotic syndrome**
  - **proteinuria**
  - **hypoproteinemia**
  - **edema**
  - **hyperlipidemia**

  Nephritic syndrome
  - **hematuria**
  - **proteinuria**
  - **impaired kidney function**
  - **hypertension**


GLOMERULAR DAMAGE
↑ Permeability of Glomerular Capillaries to Protein
PROTEINURIA (≤ 3.5 g/24 hr)
HYPOPROTEINEMIA (Albumin < 3 g/100 ml)
↓ Plasma oncotic pressure
Compensatory synthesis of proteins (including lipoproteins) by liver
↓ Plasma volume
Fluid escapes into tissue
↓ GFR
HYPERLIPIDEMIA
↑ Aldosterone secretion
Fluid retention
EDEMA

Nephrotic syndrome

- **“Primary” disease**
- **Secondary to systemic diseases**
  - **Diabetic nephropathy**
  - **Amyloidosis**

Minimal changes nephropathy (MCD)

- Most usual cause of nephrotic syndrome
- Mostly chidren
- ”Normal” histology
- Loss of foot processes

Focal segmental glomerulosclerosis (FSGS)

- **Primary**
- **Secondary**
  - **(often milder nephrotic syndrome)**
  - **extreme obesity**
  - **reflux nefropathy/ single kidney**
  - **renal artery stenosis**
  - **HIV- nefropathy**

Diabetic nephropathy

- **NIDDM - type II diabetes**

  40-50% affected after 20 years disease
- **hypertension, metabolic risk factors**

  ”nephrosclerosis”
- **as in type I diabetes**

Amyloidosis

- **Systemic - AL - 'primary'**

  monoclonal Ig light chain

  plasma cell diseases - myeloma

- **(Reactive) systemic -AA - 'secondary'**

  prealbumin

  chronic infl dis - RA, Crohn, tbc, chronic bronchitis

- **FAP - ATTR - 'Skelleftesjukan'**

  transthyretin- gen variant

Nephrotic syndrome
work up

- **Biopsy - compulsory**
- **Systemic disease?**

  SLE? Diabetes? Myeloma?
- **Malignancy?**

  Lymphoma? Cancer?

Nephritic syndrome

- **Proteinuria - ’nephrotic’**
- **Hematuria - micro~/macro~**
- **Hypertension**
- **Decreased GFR**

Glomerulonephritis

- **Acute GN**
  - **Post streptococcal nephritis**
- **Rapidly progressive GN (crescentic nephritis)**
  - **Anti-basalmembrane-nephritis (anti-GBM-nephritis)**
  - **Postinfectious crescentic nephritis**
  - **Idiopathic ~(ANCA-GN)**
- **Chronic GN**
  - **Membranous nephropathy - "membranous GN"**
  - **(FSGS)**

Poststreptococcal glomerulonephritis PSGN

- unusual in Sweden
- "nephritogenic" Gr A streptococcal inf
- tonsillitis, scarlatina, impetigo
- mostly subclinical, but…
- late effects in heart, joints, brain

RPGN

	IFL	Antibodies
• Anti-GBM-nephr 2-20%	linear	anti-GBM against Goodpasture ag
• Postinfect immune complexes 15-50%	granular	
• Idiopathic ANCA	neg	p-ANCA/c-ANCA pauci-immune

RPGN

Renal ltd disease	Systemic disease
• Anti-GBM-nephritis	Goodpasture's syndrome
• Immune complex nephritis	SLE
• Idiopathic RPGN	Wegener / small vessel vasc.

IgA -nephropathy

- **Berger, 1968 - "Berger-nephritis"**
- **most common nephritis in the world**
- **20-30 years age**
- **3-6 x more men**

IgA -nephropathy

- **PAD**
  mesangial proliferation
  ⇨ mesangioproliferative GN
- **IFL**
  mesangial IgA deposits

Systemic diseases with IgA -nephropathy

- **Henoch-Schönlein purpura**
- **ulcerative colitis, Mb Crohn, celiac disease**
- **dermatitis herpetiformis**
- **liver diseases**

SLE-nephritis

- **WHO classification based on biopsy findings:**
-
- **class I ⇒ normal glomeruli**
- **class II ⇒ mesangial GN**
- **class III ⇒ focal segmental proliferative GN**
- **class IV ⇒ diffuse proliferative GN ± ev crescents)**
- **class V ⇒ membranous nephropathy**
- **(klass VI ⇒ glomerulosclerosis - ”endstage")**
- **IFL ⇒ "full-house"**

Tubulointerstitial diseases

- **Infectious**
  - **Pyelonephritis (TIN)**
  - **Endemic nephropathy**
- **Non-infectious**
  - **Allergic TIN**
  - **Analgesic/toxic nephropathy**
  - **Gout nephropathy**
  - **Nephrocalcinosis**
  - **Granulomatous disease**

ACUTE PYELONEPHRITIS

- **ETIOLOGY** — **Bacterial infection (E. coli 80%)**
- **PATHOGENESIS** — **Ascending infection**
- **PATIENTS at RISK**
  - **a) KAD**
  - **b) Obstructive uropathy**
  - **c) Reflux**
  - **d) Old or pregnant women**
  - **e) Diabetes**
  - **f) Malformation**

CHRONIC PYELONEPHRITIS

- **ETIOLOGY** **Chronic infection?**
- **PATHOGENESIS** **Obstruction or reflux**
- **PRESENTATION** **Hypertension, progressive renal failure**
- **MICRO** **Interstitial fibrosis, tubular atrophy, FSGS**

Hemorrhagic fever with renal syndrome

NEPHROPATHIA EPIDEMICA

- EPIDEMIOLOGY — Most cases in Northern Scandinavia
- ETIOLOGY — Puumala virus(Gen Hantavirus;Fam. Bunyaviridae) in rodents (bank vole [skogssork])
- PRESENTATION — Fever, malady, acute renal failure, hematuria
- MICRO — Hemorrhagic TIN

TOXIC ATN

- ETIOLOGY — Heavy metals
Fungal toxins
Drugs
- PATHOGENESIS — Necrosis of epithelium in proximal tubuli
- MICRO — Normal BM

Ischemic Nephropathy (ATN)

- **ETIOLOGY** **Major injury/trauma hypovolemia/ hypoperfusion**
- **PATHOGENESIS** **Necrosis of tubular epithelium (distal and/or proximal)**
- **MICRO** **Damaged BM. Inflam, cylinders**

VASCULAR DISEASES

Renal damage causes hypertension, hypertension causes renal damage

- Nephrosclerosis benign/malignant
- Renal artery stenosis
- Infarction
- Vasculitis

BENIGN NEPHROSCLEROSIS

- ETIOLOGY / PATHOGENESIS
  - Primary hypertension (90-95%)
  - Secondary hypertension
- MACRO Small kidneys, thin cortex
- MICRO Hyaline arteriolosclerosis
- PREVALENS In all older people but more pronouced if hypertension

MALIGNANT NEPHROSCLEROSIS

- ETIOLOGY — Malignant hypertension
- PRESENTATION — Sympt fr other organs, hematuria, rapidly progressing renal failure
- MICRO — Fibrinoid necrosis in arterioli, thrombotic microangiopathy

RENAL ARTERY STENOSIS

- ETIOLOGY Atherosclerosis
  Fibromuscular dysplasia

- PATHOGENESIS causes renin... Low perfusion high level of

- TREATMENT PTRA

RENAL INFARCTION

- ETIOLOGY Arterial embolization
  ("cholesterol emboli")
- MACRO Wedge shaped pallor

- SYMPTOMS Sharp flank pain
  Hematuria

HYDRONEPHROSIS

- (Obstructive uropathy)
    - Dilatation of pelvis and calyces, atrophy of parenchyma
- **ETIOLOGY** High pressure
    - Congenital
    - Acquired

RENAL STONES

Urolithiasis Nefrolithiasis

Urethrolithiasis

Cystolithiasis

- ETIOLOGY
  - Increased urinary excretion of salt
  - Lack of inhibitory substances
  - Residual urin
  - Infection
  - pH

Ca-OXALATE STONES

70 % of all stones in Sweden

Hypercalcemia / -uria (10%).

Idiopathic hypercalcuria (50%)

Normal Ca in urin (20%).

Hyperoxaluria (eg bowelshuntop)

Hyperuricemia (20%)

MAGNESIUM AMMONIUM PHOSPHATE STONES (5 - 10 %)

- ”Staghorn calculus”
- Infection with urea splicing bacteria

URIC ACID STONES

- Only 25% has gout

CYSTINE STONES

- Hereditary cystinuria

TUMOURS of the KIDNEY

- BENIGN: Adenoma (papillary)
  Oncocytoma
  Angiomyolipoma

- MALIGNANT: Renal cell carcinoma

RENAL CELL CANCER

2% of all cancer, 1000 cases/year in Sweden

men : women = 11 : 7

elderly people (>70 yo)

50 % incidental finding!

- ETIOLOGY Smoking, obesity, high blood pressure, diabetes mellitus
- HEREDITY < 5 % (von Hippel-Lindau)
- CLIN FEAT Hematuria
  painful tumour mass
  - anemia
  - high SR
  endocrine activity

MICRO Clear cell (= ”conventional”)
Papillary
Chromofobe
Collecting duct

TREATMENT Surgery
PROGNOSIS 50% 5 years
-No met. 70 %
-Tumour in vein 15%

PROGNOSTIC FACTORS

GRADING

four grades according to Fuhrman

STAGING

pTNM by UICC 2002