• 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

↓ Plasma volume

Compensatory synthesis of proteins (including lipoproteins) by liver

Fluid escapes into tissue

↓ GFR

Hyperlipidemia

↑ Aldosterone secretion

Fluid retention

↓ GFR

EDEMA
Nephrotic syndrome

• “Primary” disease
• Secondary to systemic diseases
  – Diabetic nephropathy
  – Amyloidosis
Minimal changes nephropathy (MCD)

- Most usual cause of nephrotic syndrome
- Mostly children
- ”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 - 'Skelleftešjukan'**
  
  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)
  - Membranoproliferative GN (dense deposit
Poststreptococcal glomerulonephritis (PSGN)

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

IFL Antibodies

- Anti-GBM-nephr linear anti-GBM against Goodpasture
  2-20% ag

- Postinfect granular immune complexes
  15-50%

- Idiopathic 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 × more men
IgA nephropathy

- PAD
  mesangial proliferation
  \[\Rightarrow\] 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 pronounced 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  Low perfusion
           causes high level of renin...

• 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)
Hyperuricicemia (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
  - 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