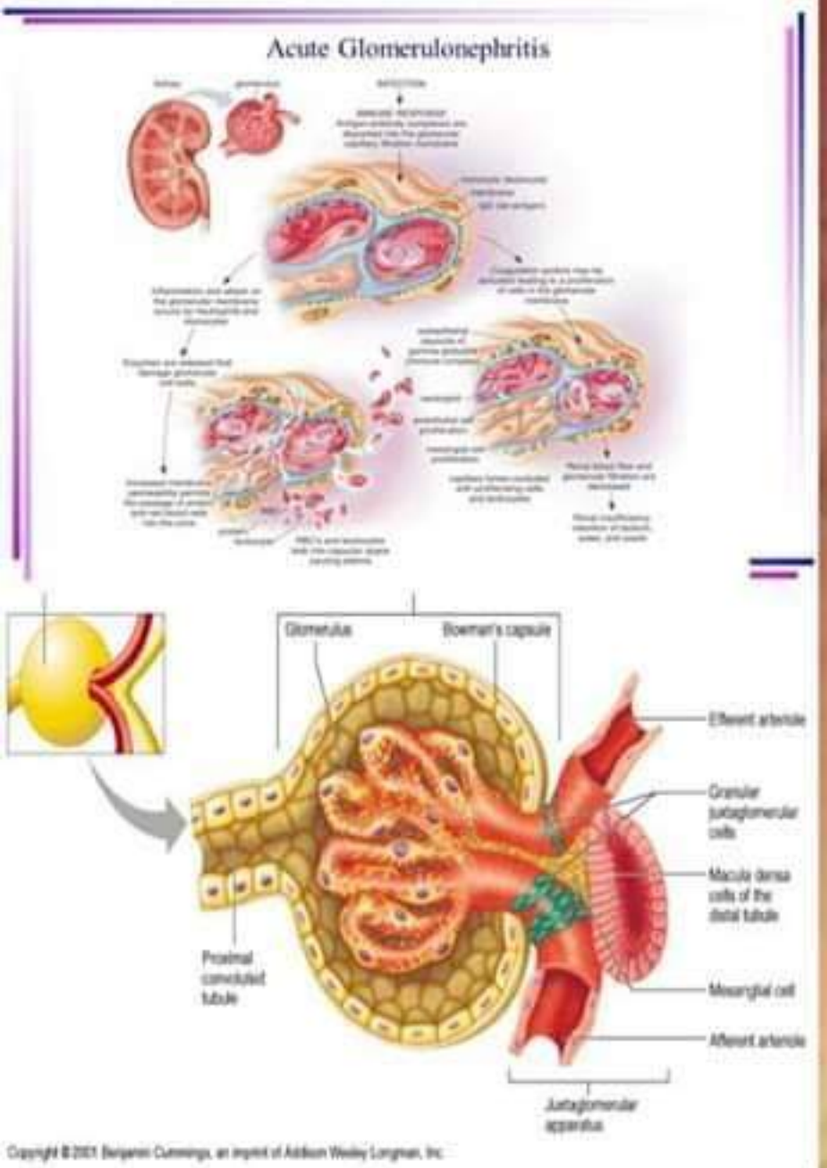
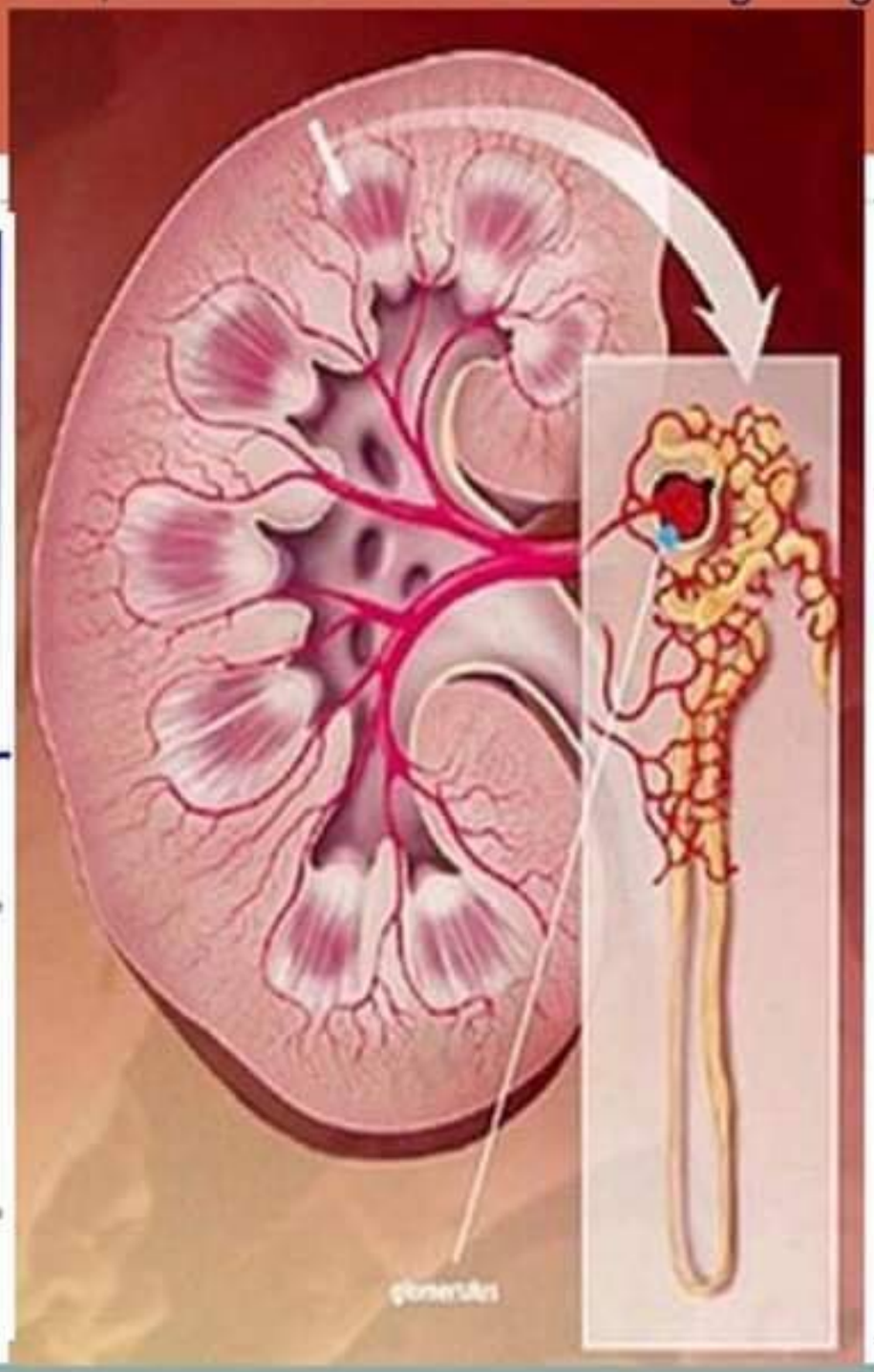
Acute glomerulonephritis





Objectives

- Definition
- Classification
- Acute streptococcal GN
- IgA nephropathy
- Membranoproliferative GN
- Henoch Schonlein Purpura
- Lupus Nephrits
- Alport syndrome

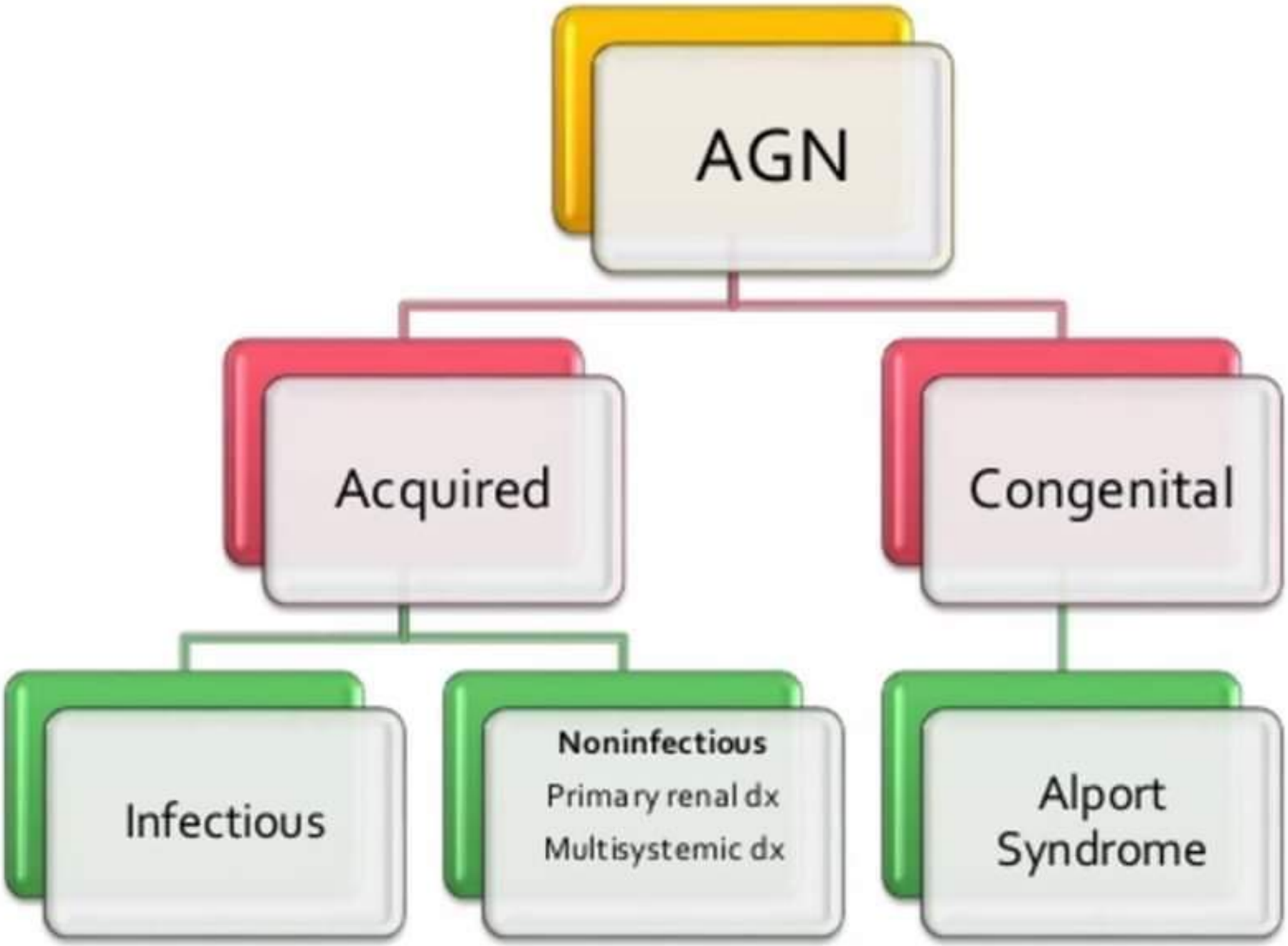
Glomerulonephritis (GN)

Glomerulonephritis is both a clinical and histopathologic term signifying inflammation and proliferation of the glomerulus

Severe glomerular histopathologic and clinical entities, such as

- anti-GBM antibody disease(Goodpasture syndrome)
- Wegener granulomatosis
- Idiopathic and Rapidly progressive GN
- May be considered as differential diagnosis of GN, but are exceedingly rare in children.

Classification



Classification of GN - Acquired

1. Primary glomerulonephritis
   - Immune complex GN
     - Post infectious glomerulonephritis
     - IgA nephropathy(Berger disease)
     - Membrenoproliferative glomerulonephritis
     - Membranous GN (idiopathic)
   - Anti-GBM antibody mediated GN
   - Uncertain etiology- Minimal change GN, Focal segmental GN

Classification of GN

Systemic

2. GN associated with systemic disorders
   - Immunologically mediated
     - Henoch-Schonlein purpura
     - SLE and other collagen diseases e.g. scleroderma
     - Vasculitides: polyateritis nodosa, wegener's granolomatosis
     - Systemic infections (subacute bacterial endocarditis, shunt nephritis, syphilis, malaria, hepatitis B, HIV)
3. Hereditary disorders- Alport Syndrome

Clinical patterns of GN

1) Asymptomatic hematuria and / or proteinuria
2) Acute nephritic syndrome
   - Hematuria
   - Oliguria
   - Hypertension
   - Edema
3) Nephrotic syndrome
   - Heavy proteinuria
   - Hypoalbuminemia
   - Edema with hyperlipidemia

Acute Post streptococcal Glomerulonephritis

- Follows a recent history (7-14days previously) of group A -β hemolytic streptococcal infection involving the **pharynx and the skin(impetigo)**

  - **GFR= 38* ht (cm)/ plasma creatinine (umol/l)**

- Clinical presentation
  - Gross hematuria coffee/tea colored
  - Edema (periorbital,facial and extremities) caused by sodium and water retention
  - Hypretension * headache
  - Oliguria
  - Azotemia- *an increase in BUN and Creatinine related to a decrease in GFR*
  - Fever is uncommon

APSGN

- Investigations
  - Positive culture (throat swab)
  - Elevated Anti-streptolysin O titer
  - High titers of other antistreptococcal antibodies
  - Microscopy- dysmorphic RBC, heme granular casts, pyuria, white cell
  - Hypocomplementemia esp. C_3, C_4
  - CXR- cardiomegally, pleural effusion, pulmonary edema
  - Increase in serum creatinine
  - Severe glomerular injury may be accompanied by massive proteinuria (nephrotic)
- RENAL BIOPSY
  - In persistent hypocomplementemia and nephrotic syndrome

Treatment and management

Supportive, depends on the degree of renal impairement and hypretension

- Antibiotic therapy (penicillin 10day course) if infection is still present
- Diuretics
- Antihypertnsives
- Close monitoring and reduction of salt and water intake
- In severe renal failure : hemodialysis or peritoneal dialysis
- Corticosteroids may also be administered

- Good prognosis- resolves in 2-3 weeks

IgA Nephropathy

- **Characterised by IgA deposition in the mesangium**
- Classic presentation is asymptomatic gross hematuria during acute unrelated illness
- IgA is elevated in most cases.
- complement is not depressed
- Gross hematuria resolves within days, there is no serious sequelae in 85% of cases

IgA Nephropathy

Investigations

- Hematuria- exclude other causes (SLE- ANA, Dnase, ASOT)
- Renal biopsy- presence of mesangial IgA, immunofluorescent deposits with lesser amounts of IgG,IgM
  - Light microscopy – mesangial proliferation and increased mesangial matrix

Treatment

- is not indicated and prognosis is good in most cases
- In severe proteinuria, hypertension, or renal insufficiency – corticosteroids, immunosuprressive drudg and omega-3 from fish oil are helpful

Henoch Schonlein Purpura

- Most common small vessel vasculitis in childhood
- It is accompanied by arthritis, abdominal pain, bloody diarrhea
- Approximately 50% of patients develop reanl mainfestations, which vary from asymptomatic microscopic hematuria to severe, progressive glomerulonephritis

pathogenesis

Deposition of IgA in the glomerulus.

This is analogous to the IgA deposits seen in systemic small vessels, primarily those of the skin and intestine

The glomerular findings are indistigushable from those of IgA nephropathy

HSP

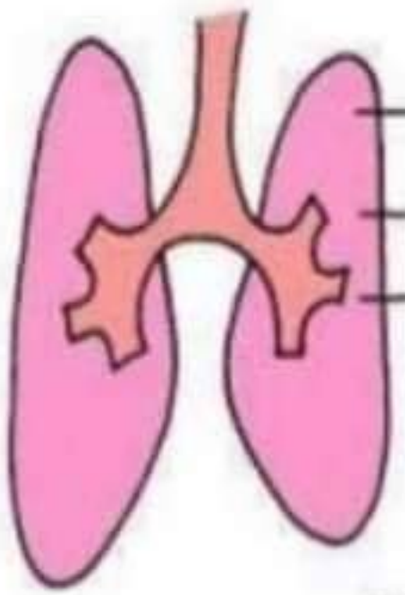
- Clinical presentation
  - Nephritis follows onset of the rash, often weeks or months atfer initial presentation of the disease
  - Nephritic syndrome picture
  - Nephritic- nephrotic picture
- Prognosis
  - In most patients its excellent- spontaneous and complete resolution occurs in those with mild symptoms
  - Some my progress to chronis renal failure

Membranoproliferative GN

- Manifested histologically by alterations of the GBM, mesangium and proliferation of the glomerular cells
- Type 1 – 80% of case
  - Characterised by subendothelial electron dense deposits of C_3 $ IgG components in an irregular manner
  - Good prognosis Responds to corticosteroids
- Type 2 – Dense Deposit disease
  - Worst prognosis, less responsive to treatment

- MPGN Type 1
  - Occurs with other diseases
    - SLE
    - Hepatitis B,C
    - Chronic liver dx
    - Chronic baterial infections

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)



- Tachypnea
- Cough
- Pleural Inflammation/ Effusion

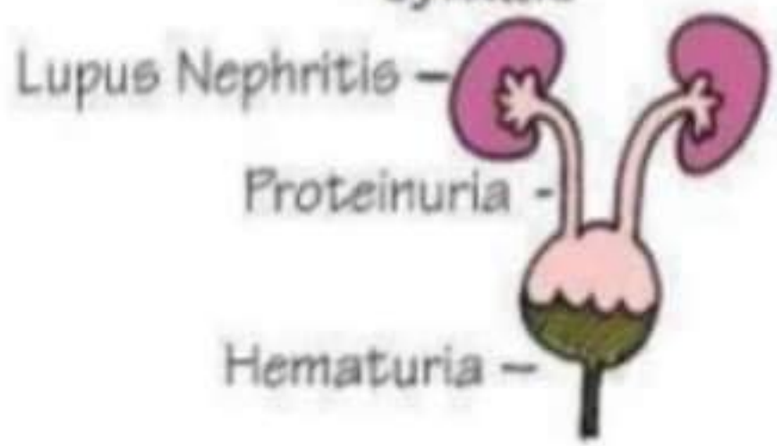
Photo Sensitivity

Erythematous Rash To Areas Exposed To Sunlight

Butterfly Rash Over Cheeks

- Weight Loss
- Fatigue
- Fever ↑Infection
- Arthritis
- Emotional Lability
- Hematologic Disorders
- Neurologic Disorders

- Raynaud's Phenomenon
- Pericarditis
- Vascular Inflammation



- Lupus Nephritis
- Proteinuria
- Hematuria

Predisposing factors -SLE

- **Geography.**
- **Race.**
- **Sex.** Affects 1 in 700 women of the child bearing age.
- **Age-** high incidence 20-29 years
- **Family history**
- **Blood group** – studie by *Baart De La Faille*

It's a disease of the young and old

Snoop's daughter has lupus

Lupus Nephritis

- renal involvement is indicated by varying degrees of hematuria and protenuria, accompanied by renal insufficiency and hypretension
- Significant renal involvement requires treatment with immunossupressive drugs and close monitoring
- ESRD develops in 10-15% of children with SLE

Hereditary GN

- Alport Syndrome
  - It is a chornic form of GN and thus does not present with the clinical features seen in patients with an acute process
  - X linked
  - Related to mutation of gene that encode heterodimers ($\alpha3,\alpha4,\alpha5$) of type 4 collagen found in
    - Cochlea (nerve deafness)
    - Eye- lens dislocation, posterior cataract, corneal dystrophy
    - Nephritis
  - ESRD occurs
  - No treatment, supoortive management for associated hypertension