SNS COLLEGE OF ALLIED HEALTH SCIENCES

SNS Kalvi Nagar, Coimbatore - 35

Affiliated to Dr MGR Medical University, Chennai

DEPARTMENT OF CARDIOPULMONARY PERFUSION CARE

TECHNOLOGY

COURSE NAME: PATHOLOGY II

II YEAR

UNIT III : PATHOLOGY OF KIDNEY

TOPIC 5 : NEPHRITIC SYNDROME

Glomerulus

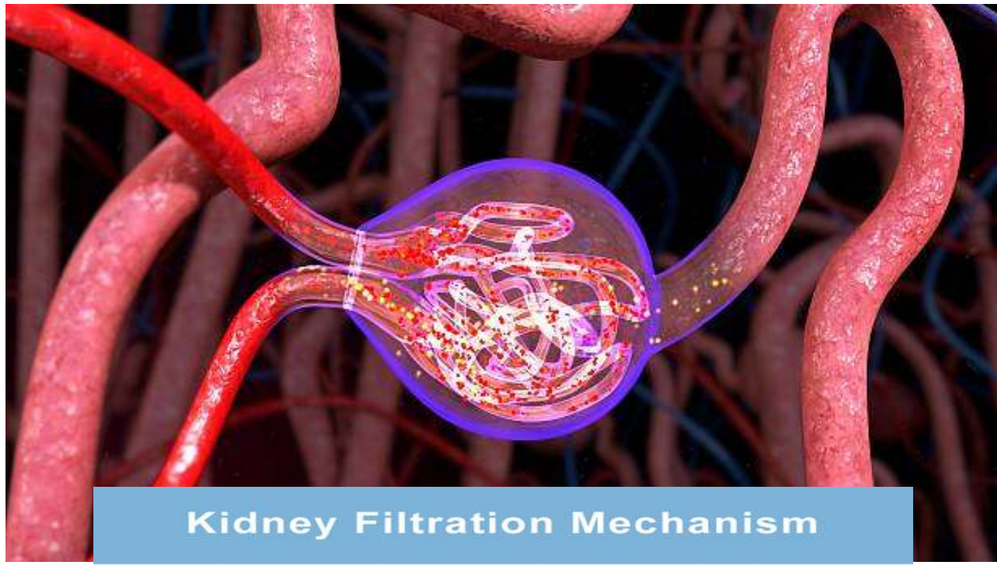
- The glomerulus is **a loop of capillaries twisted into a ball shape**, surrounded by the Bowman's capsule.

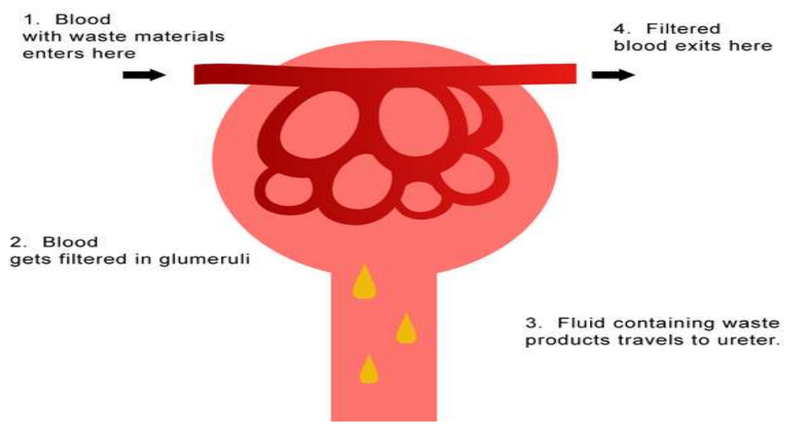
- The **glomerulus** filters the blood

- Approximately 1 million glomeruli, or filters, in each kidney.

- The glomerulus is **attached to the opening of a small fluid-collecting tube called a tubule**.

- The filtrate is captured by Bowman's capsule and directed to the proximal convoluted tubule.



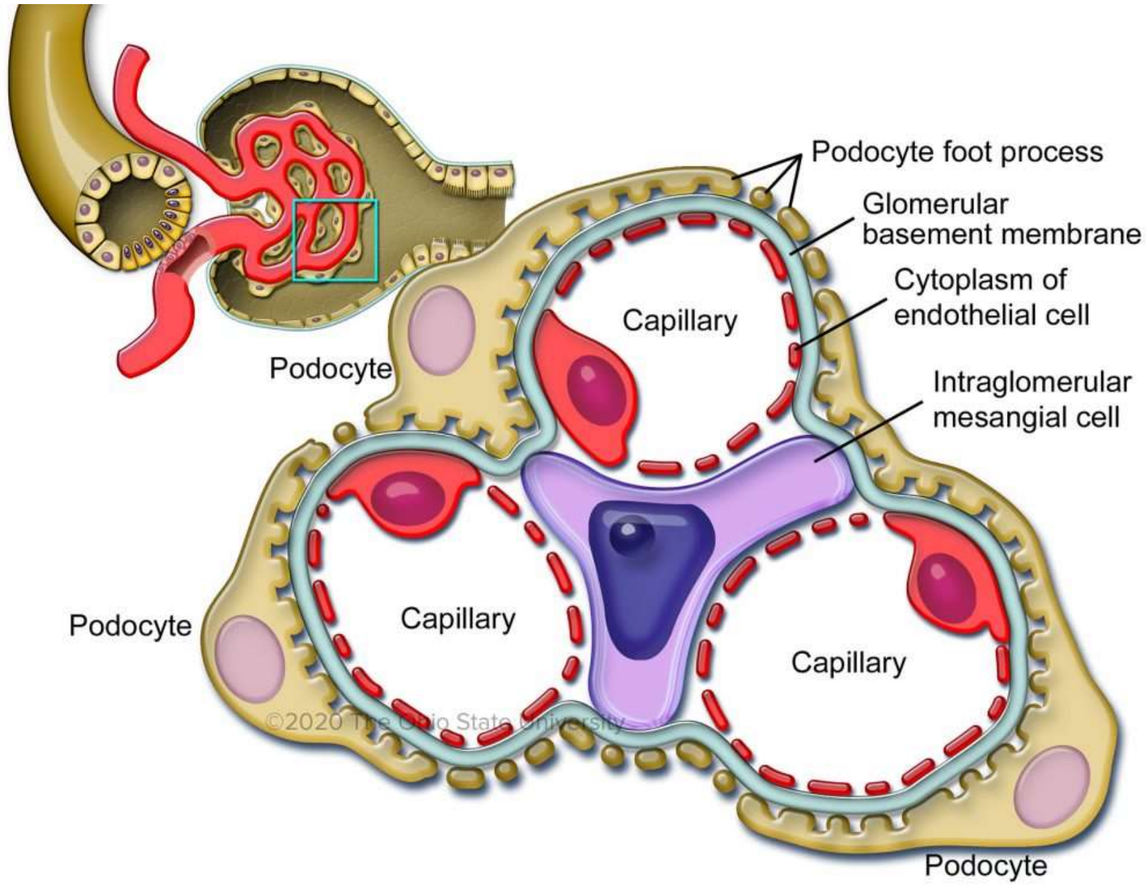


Microscopic Structure of Glomerulus

- The outermost part of glomerular capsule is a **simple squamous epithelium.**

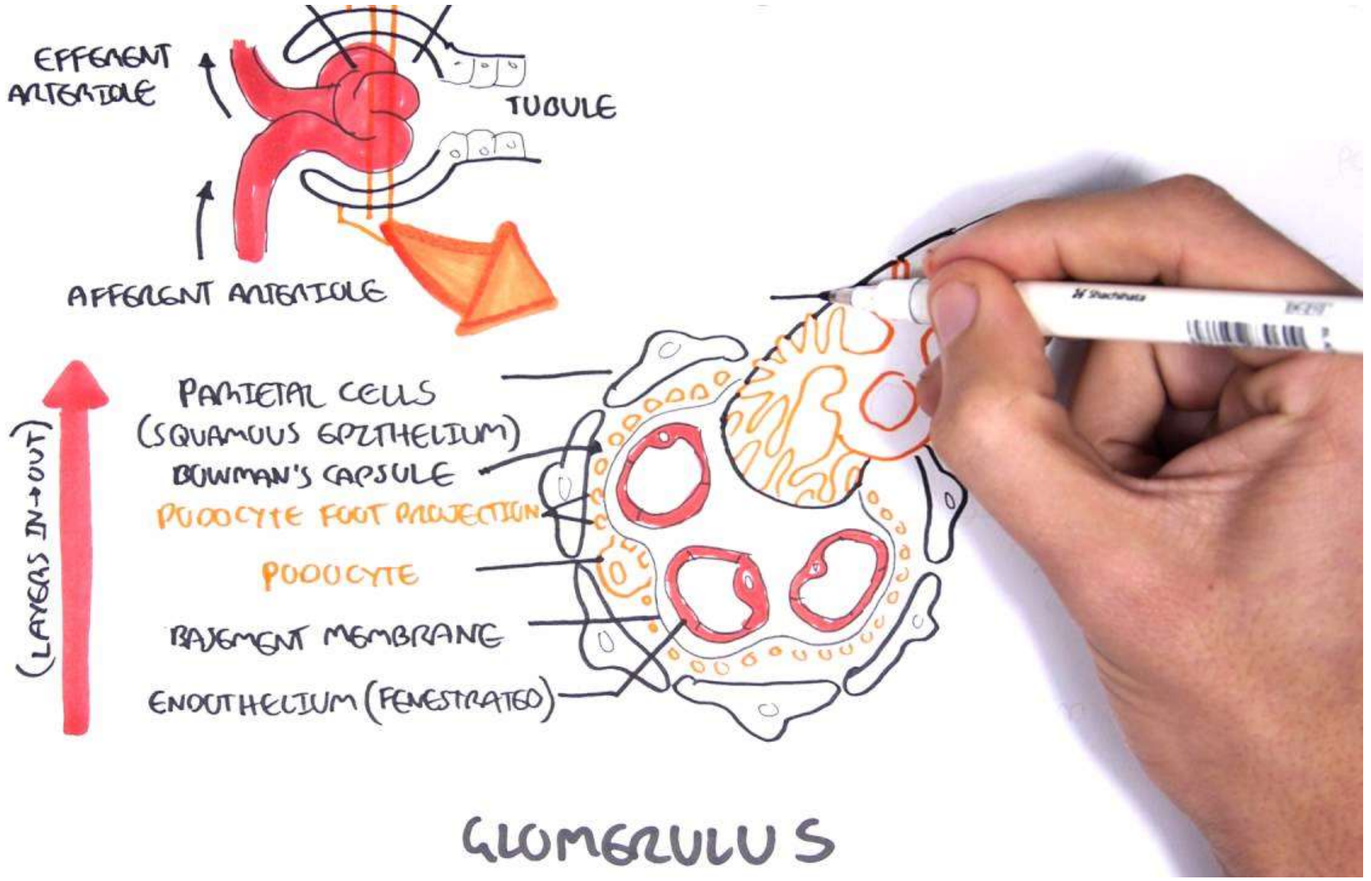
- It transitions over the glomerulus as uniquely shaped cells **(podocytes)** with finger-like arms that cover the glomerular capillaries.

- A **thin basement membrane** lies between the glomerular endothelium and the podocytes.

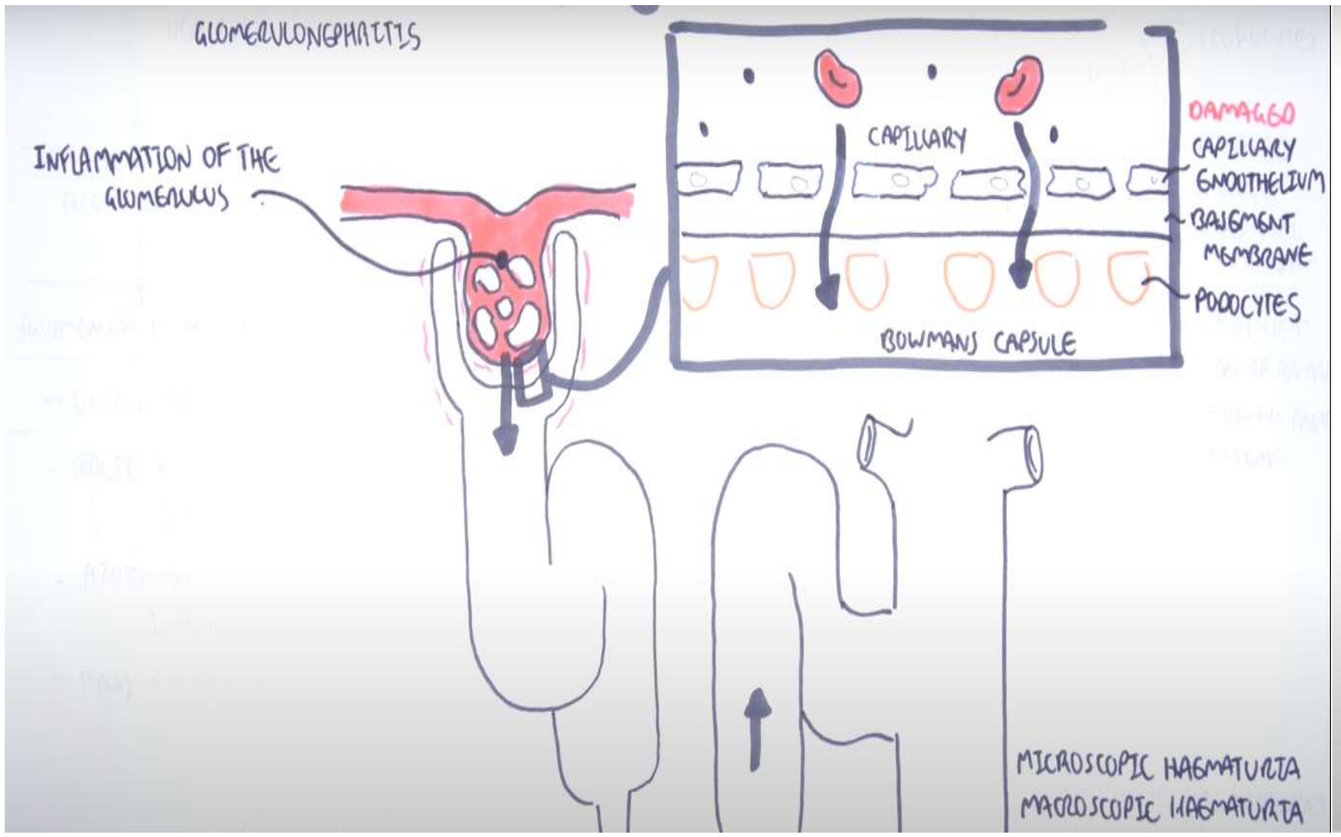


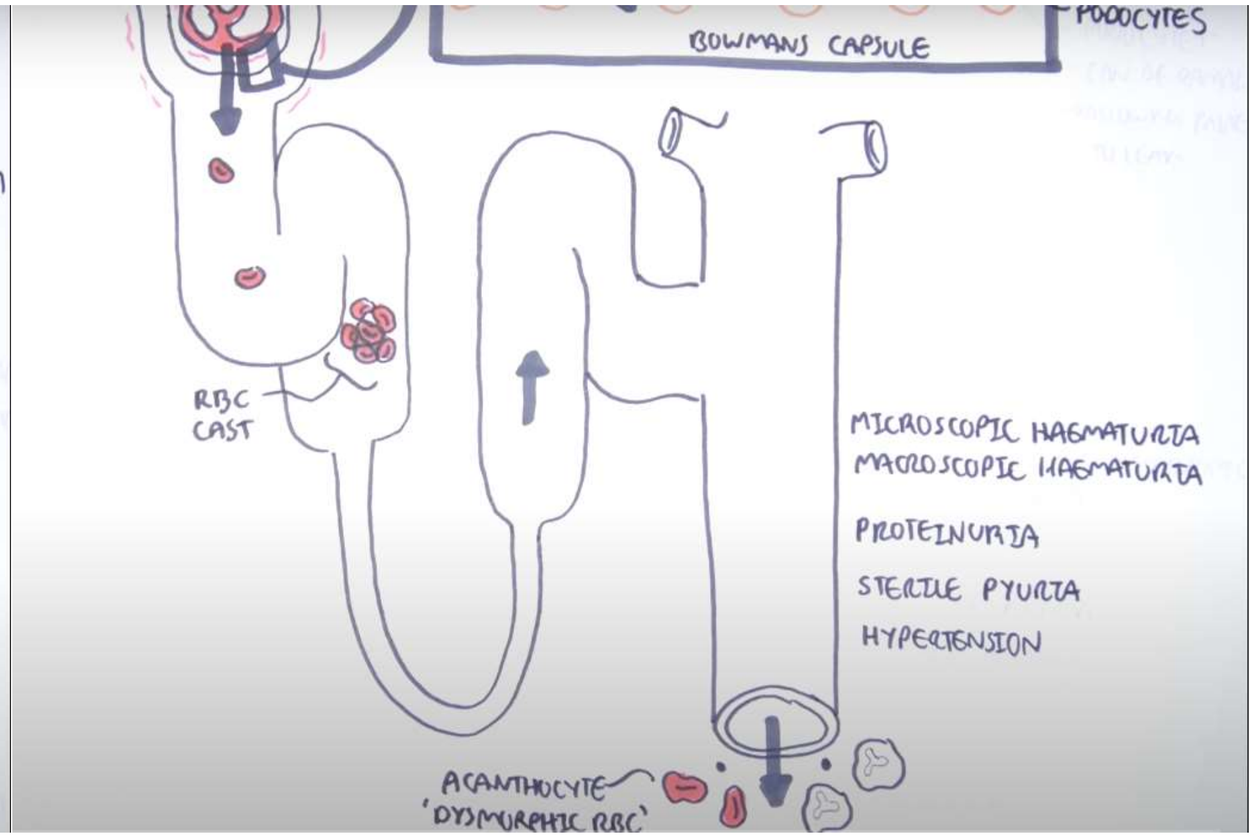
Nephritic Syndrome

Nephritic syndrome is characterized by **inflammatory rupture of the glomerular capillaries**, with resultant bleeding into the urinary space; proteinuria and oedema may be present but usually are mild.



Dysmorphic RBC





Clinical Findings

- Mild proteinuria.
- Mild edema.
- Oliguria.
- Azotemia (Increased urea and creatinine levels in blood due to decreased GFR)
- Hypertension (due to Fluid retention / increased renin secretion)
- Smoky brownish urine
- Hematuria ----------> ***Results from leakage of dysmorphic RBC directly from glomerular capillaries into the Bowman space.***

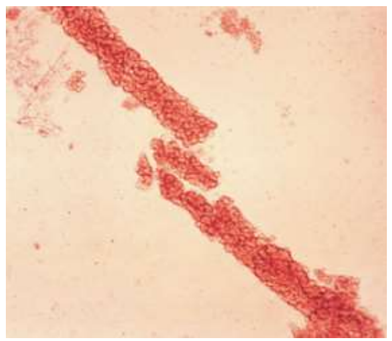
Clinical Findings

- Many of the RBC are aggregated into the shape of the renal tubules and embedded in a **proteinaceous matrix forming RBC casts** that can be observed in the urine.

- Proteinaceous is the one which cause these RBCs to adhere to each other.

- The hemodynamic changes caused by the rupture lead to a reduction in the glomerular filtration rate (GFR).

- **The RBC casts are a sign of a disease affecting the glomerulus**



Pathogenesis

- Nephritic syndrome is an immune mediated disease and this immune reaction is usually an immune complex
- The immune complex
- Creates an inflammatory reaction
- Activation of the complement by the alternative pathway
- Accumulation of c3a & c5a ---→ chemotaxis of neutrophils
- Contribution of the damage which occurred in the blood vessels
- The blood vessels inside the glomerulus react to this contribution
- Mesangial& endothelial proliferation.

Pathogenesis

Diffuse (rapid) proliferative glomerulonephritis can occur the following:

- Type 1 = Anti basement membrane anti bodies (Good pasture syndrome).
- Type 2 = Post infectious glomerulonephritis / SLE class 4.
- Type 3 = Wagener granulomatosis (Pauci-immune).

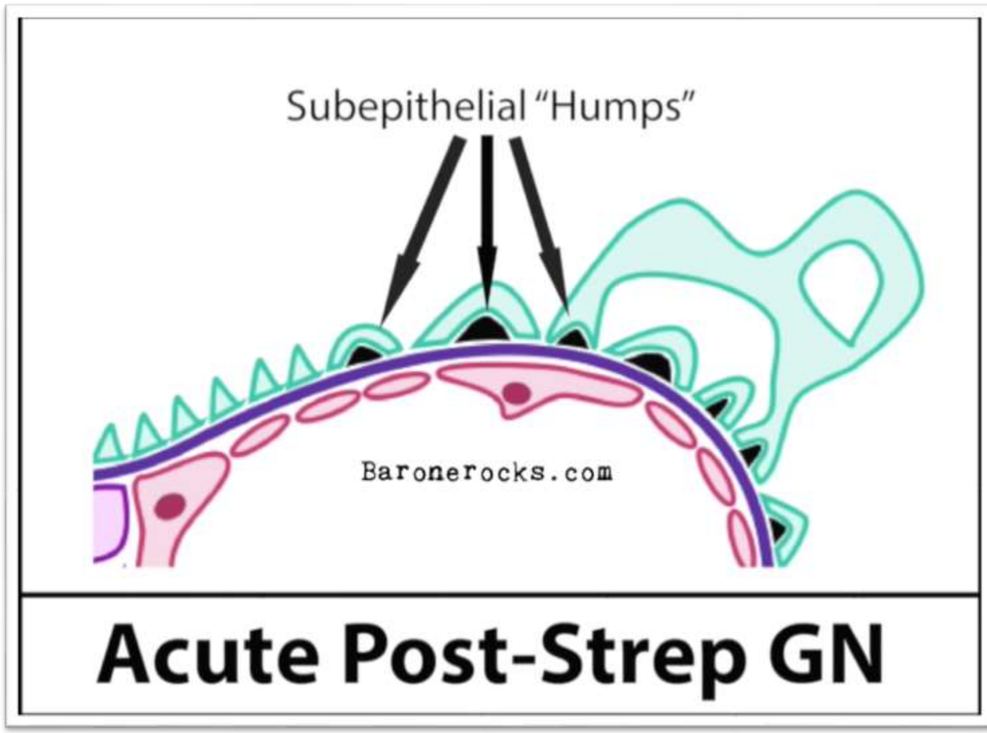
Post streptococcal glomerulonephritis

- Also called **acute post infectious proliferative glomerulonephritis.**

- It is the prototype of the nephritic syndrome.

- It is immune complex disease with the **antigen being of streptococcal origin.**

- This disease is common in young people.



It's usually occurs 1-4 weeks following upper respiratory tract infection (mainly throat infection by group A β-hemolytic streptococci)

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The child starts having edema

↓

Changing in urine color

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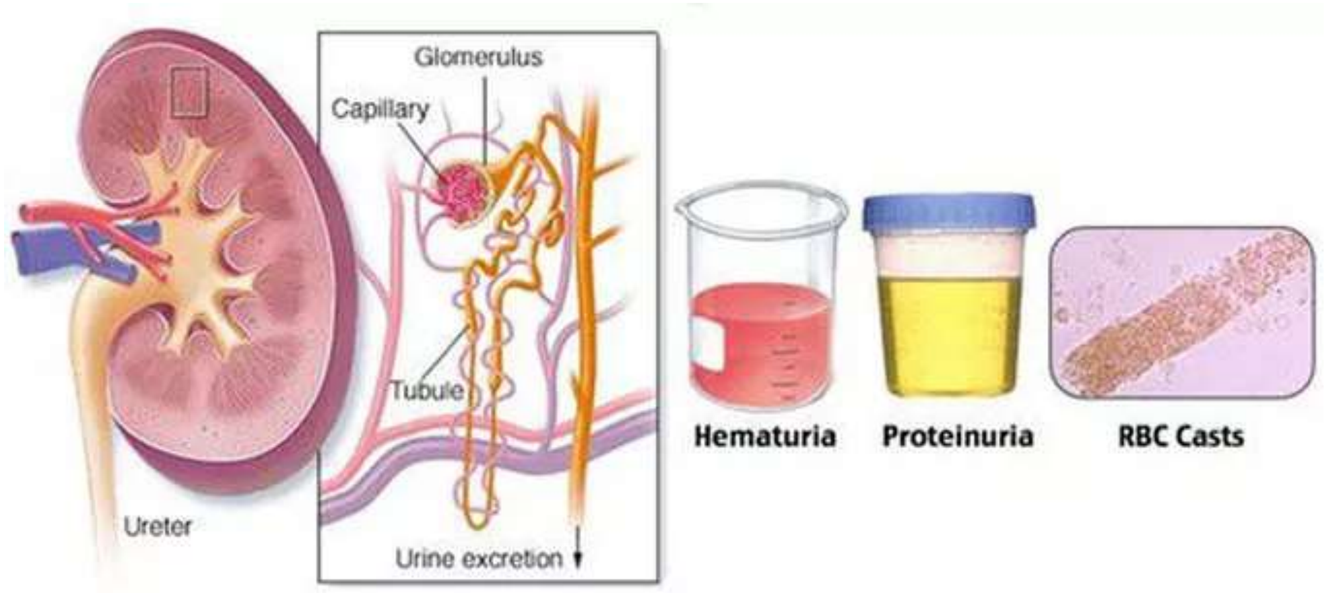
Development of nephritic syndrome.



Reactions on kidney

- Innumerable punctuate haemorrhages on the surface of both kidneys.

- Enlarged, Hyper-cellular, swollen, blood less glomeruli with proliferation of mesangial and endothelial cells

- Characteristic electron-dense "humps" on the epithelial side of basement membrane

- **High power:** There are RBCs & fibrin in the bowman's space due to the inflammatory reaction that caused damage to the basement membrane and it is leaking protein, RBC & fibrin.

- **Immunofluorescence:** There are chunks of positive immune complexes containing IgG

Alport syndrome (congenital/hereditary nephritis)

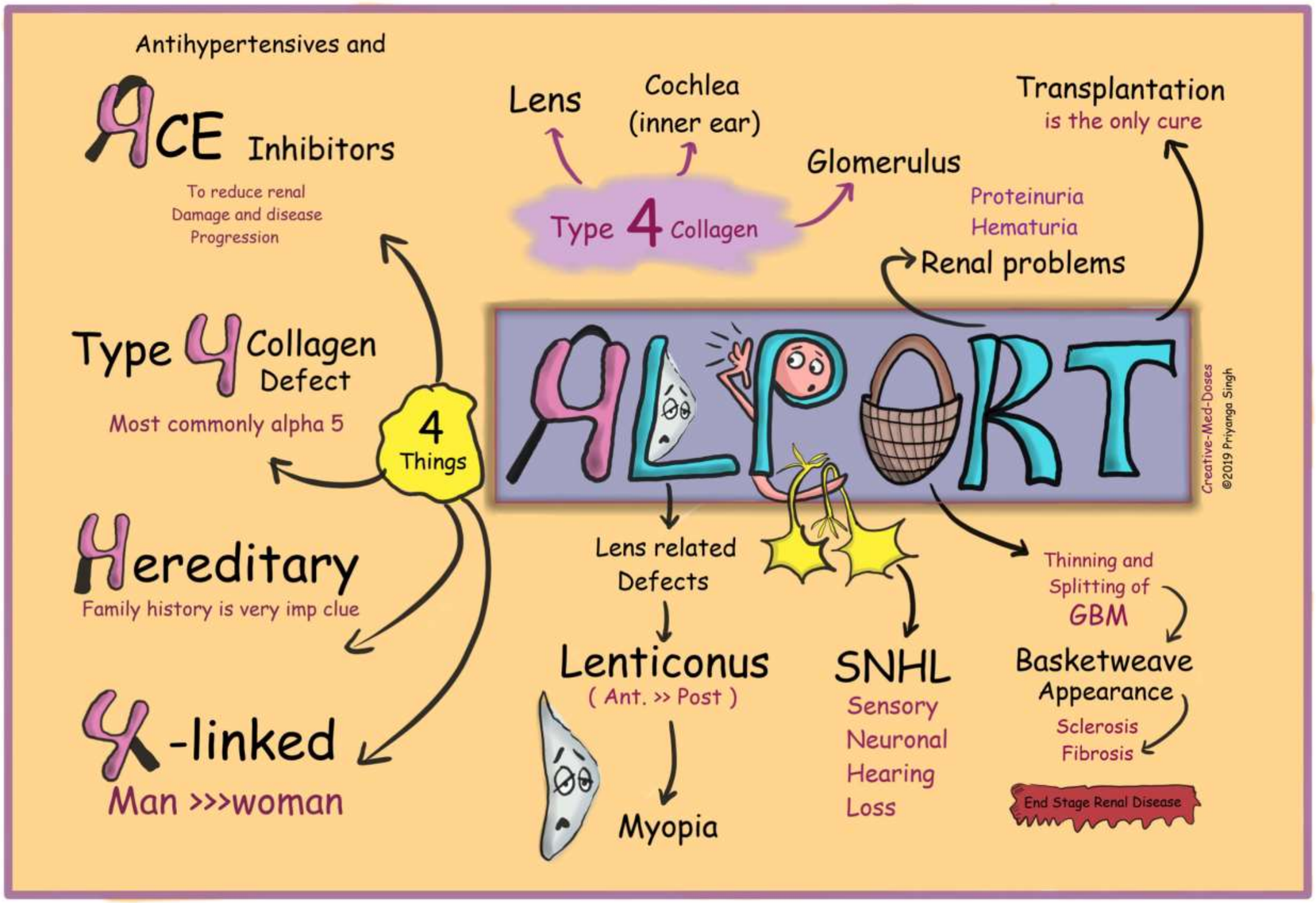
- It is a disease that usually affects children
- Split basement membrane due to hereditary nephritis

Pathogenesis:

- Caused by mutations in gene encoding for certain protein chain (α-5 chain)

Prognosis:

- Often progressing to end stage renal disease by 30 years of age.


Antihypertensives and
ACE Inhibitors
To reduce renal Damage and disease Progression
Lens
Cochlea (inner ear)
Type 4 Collagen
Glomerulus
Transplantation is the only cure
Proteinuria
Hematuria
Renal problems
Type 4 Collagen Defect
Most commonly alpha 5
4 Things
ALPORT
Creative-Med-Doses
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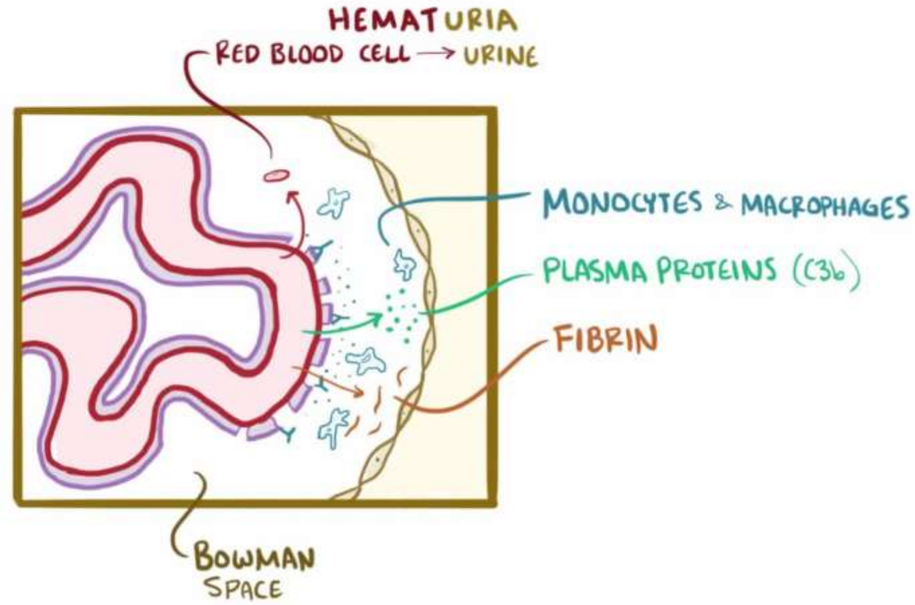
Hereditary
Family history is very imp clue
X-linked
Man >>>woman
Lens related Defects
Lenticonus
(Ant. >> Post)
Myopia
SNHL
Sensory Neuronal Hearing Loss
Thinning and Splitting of GBM
Basketweave Appearance
Sclerosis
Fibrosis
End Stage Renal Disease

Rapidly progressive (crescentic) glomerulonephritis (RPGN)

- The formation of crescents between the Bowman's capsule and the glomerular tuft

- This result from **deposition of fibrin** in the Bowman space and from proliferation of parietal epithelial cells of the Bowman capsule.

- This leads to rapid and progressive loss of renal function associated with **severe oliguria** and if untreated even death

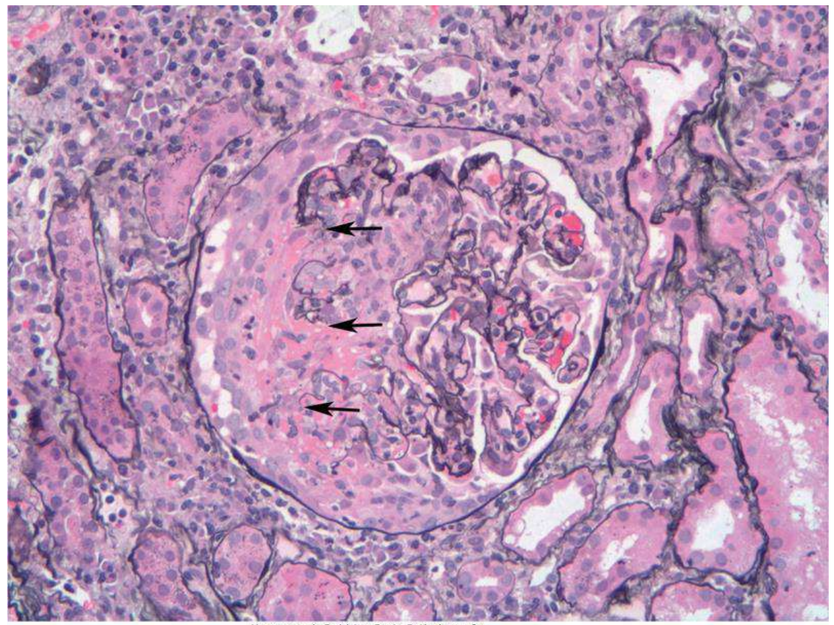


Etiology:

- Post streptococcal infection
- Anti glomerular basement membrane antibodies (non-streptococcal)

Morphology:

- Areas of necrosis with rupture of capillary loops
- Destruction of normal glomerular structures
- The adjacent crescent-shaped mass of proliferating cells
- Leukocytes filling the urinary space.



Kumar et al: Robbins Basic Pathology, 9e.
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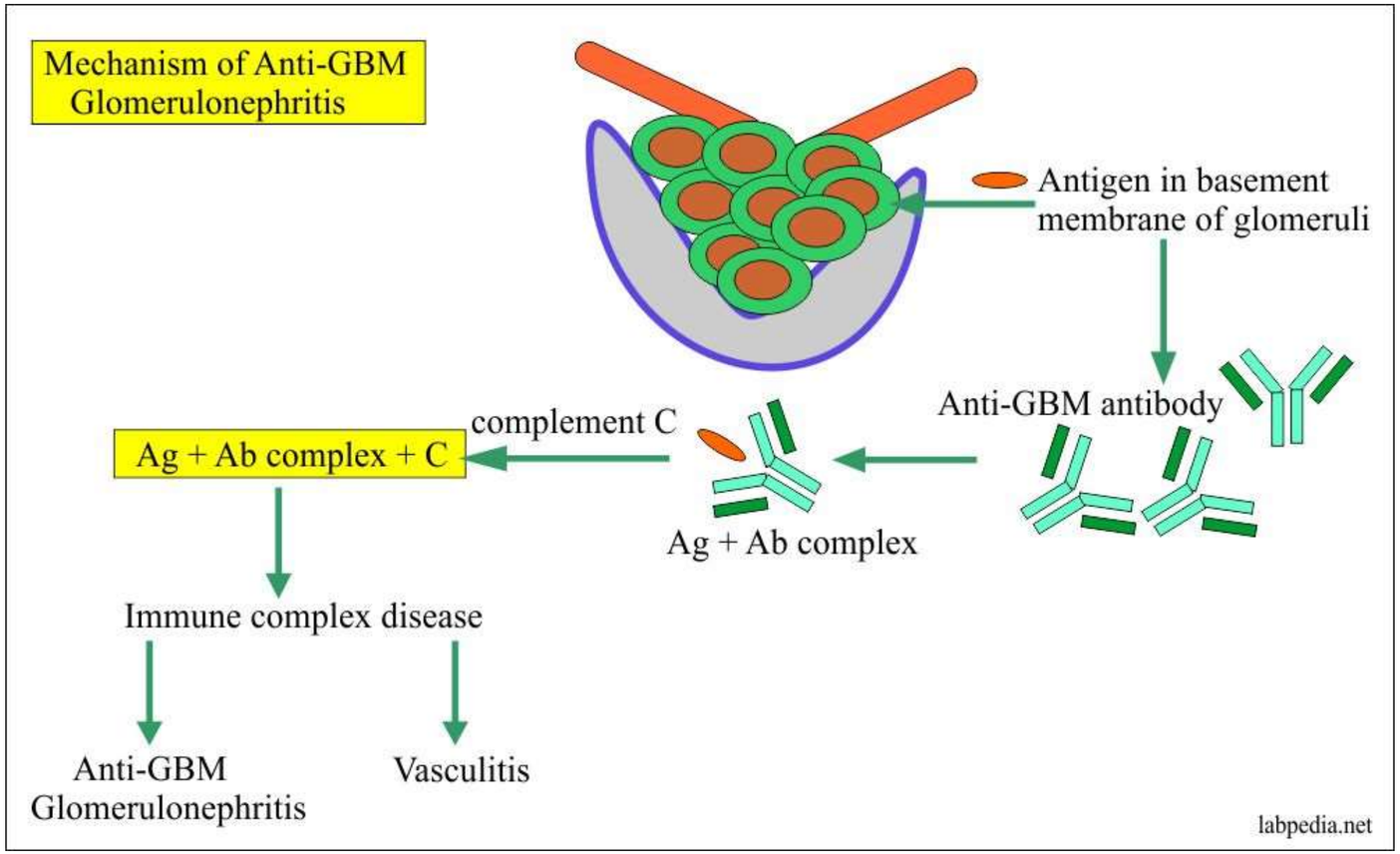
Anti glomerular basement membrane disease

- Also called **Good pasture syndrome**

- It is caused by the formation of antibodies **(anti glomerular basement membrane antibodies)**, which are directed against antigen in the glomerular and pulmonary alveolar basement membranes.



- It usually affects young males.
- It can be characterized by the **formation of crescent**. (The crescent consists of parietal epithelial cells with some inflammatory cells)

Proliferation will lead to **rupture of the basement membrane**

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Rbc’s & fibrin goes to bowman's space

↓

Reactive proliferation of parietal cells

↓

They will form the **crescent**

Wagener granulomatosis

- Also called **Pauci-immune glomerulonephritis**
- It is an auto immune disease, that causes inflammation of blood vessels (vasculitis)
- It is caused by autoantibodies against **proteinase 3**
- **Granulomas and patchy necrosis** in arteries and veins
- Increased **Anti-neutrophil cytoplasmic antibodies (ANCA)** is seen.

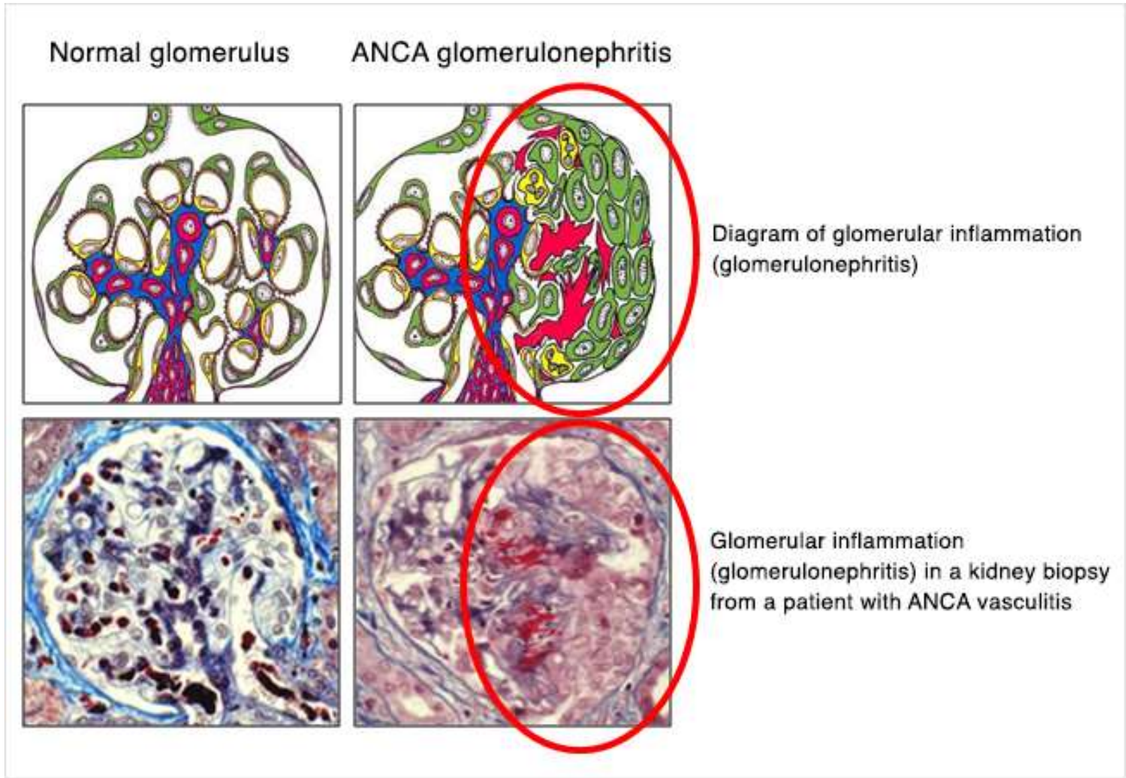


Diagram of glomerular inflammation (glomerulonephritis)

Glomerular inflammation (glomerulonephritis) in a kidney biopsy from a patient with ANCA vasculitis

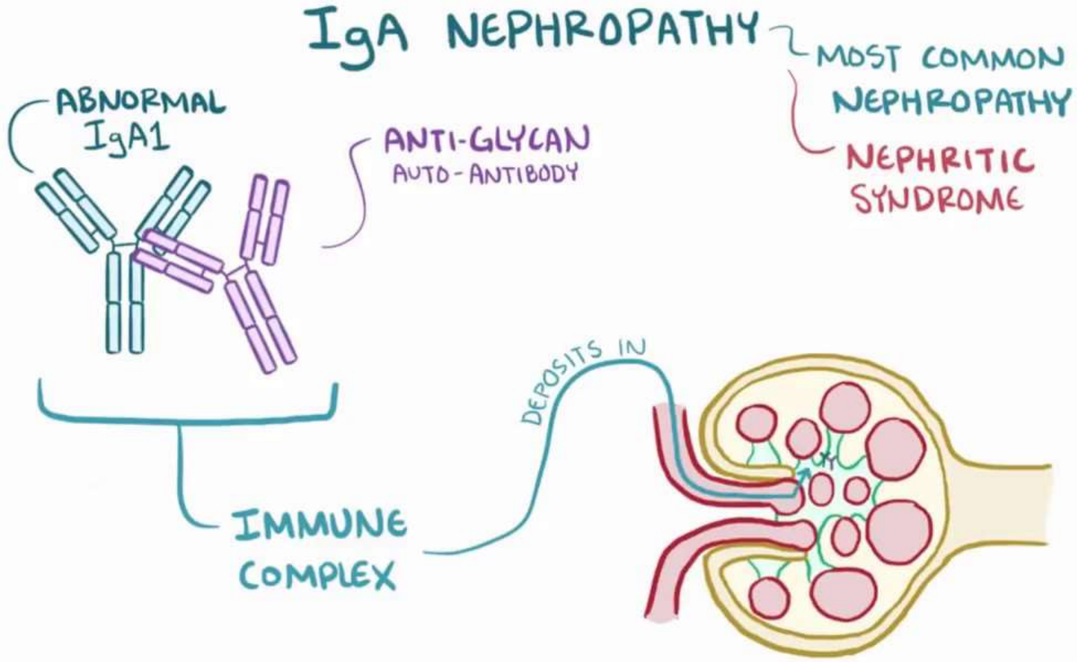
IgA Nephropathy

- Also called **Berger disease**

- It is **the most common cause of nephritis** in worldwide

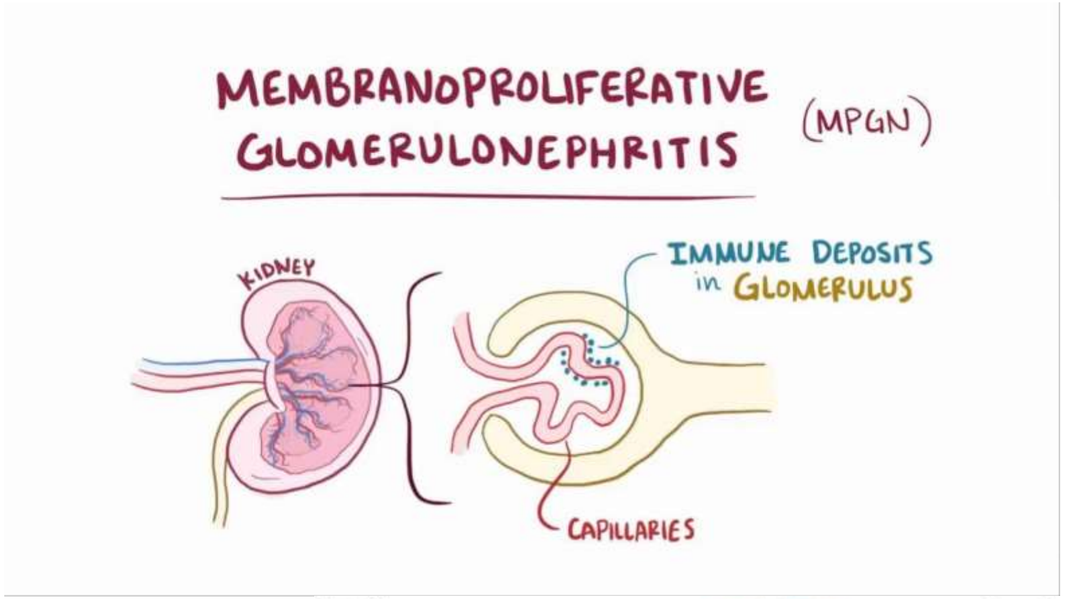
- **IgA nephropathy** is characterized by benign **recurrent hematuria** in children (only microscopic hematuria), usually following an **infection**, lasting 12 days, and usually of **minimal clinical significance.**

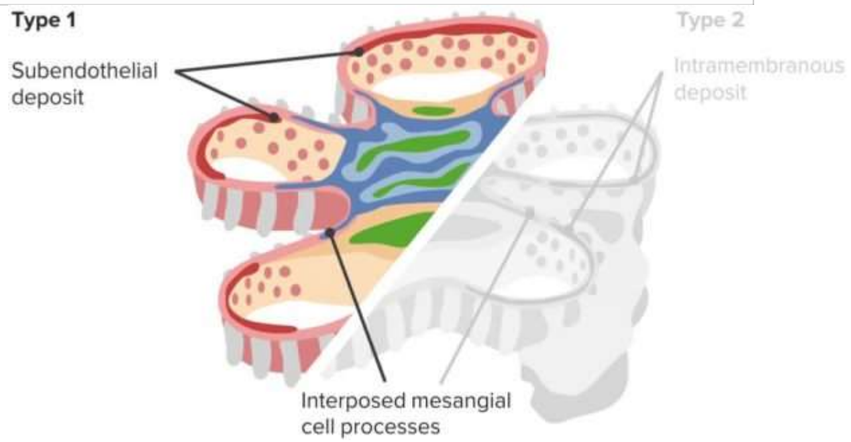
- It is also characterized by over production of **IgA**



Membranoproliferative glomerulonephritis

- Endocapillary proliferation, (endothelial cells are proliferated)
- **Clinical characteristics:** include slow progression to chronic renal disease.
- **Histological characteristics:** include both basement membrane thickening and cellular proliferation.
- There is **splitting in the glomerular basement membrane**.
- **It affects the endothelial & mesangial cells.**





Investigations

- Arterial Blood Gases
- BUN
- Blood Chemistry
- Complete Blood count
- Kidney Biopsy
- Urinalysis

Management

- Antibiotic Penicillin
- Corticosteroids and immunosuppressive drugs
- Dietary protein
- Sodium protein
- Loop diuretics and Anti-hypertensive to control hypertension