- 10 year old boy presented with hematuria. He had symptoms of an upper respiratory tract infection a few days back. He gave a history of similar episode in the past which later subsided.

- O/E: elevated blood pressure.

- Urine examination: Protein +

  RBC +++

IgA NEPHROPATHY

- Most common cause of hematuria (gross & microscopic)

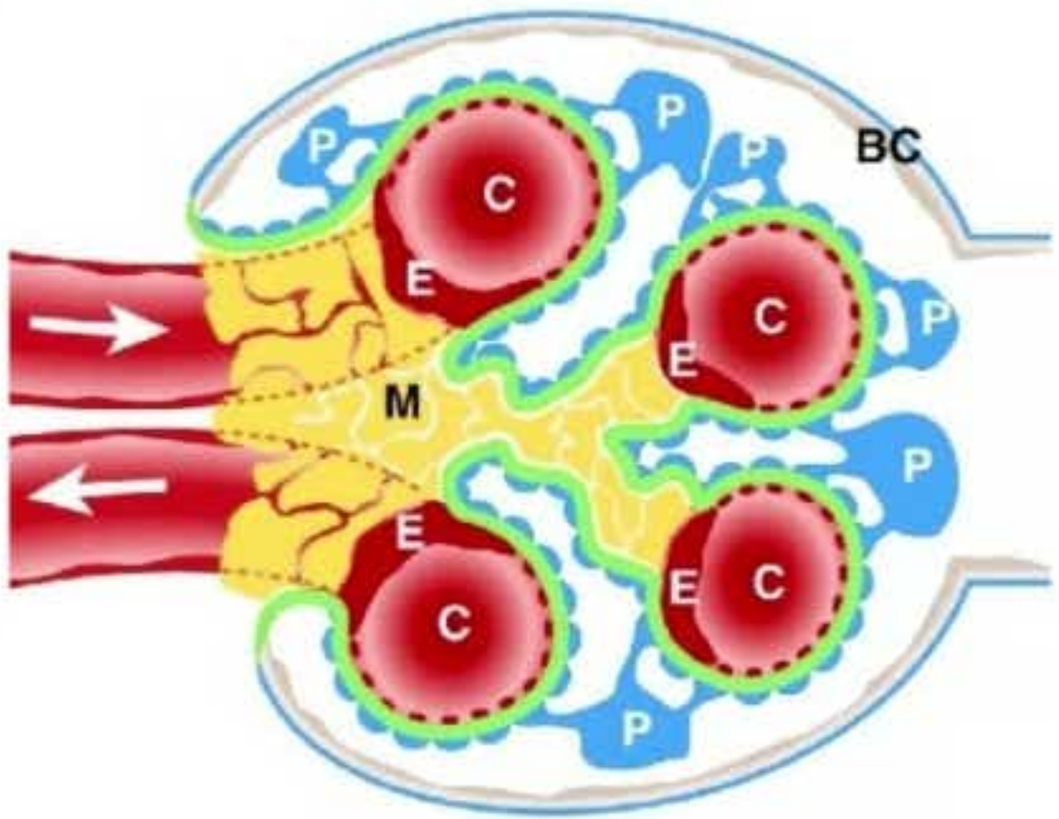
- Most common glomerular disease worldwide

- Children & young adults

- Following URTI / GIT infections/ UTI

- **HALLMARK**

"Deposition of IgA in mesangium"



Bowman's Capsule

Capillaries

Endothelial cells

Mesangium

Podocytes

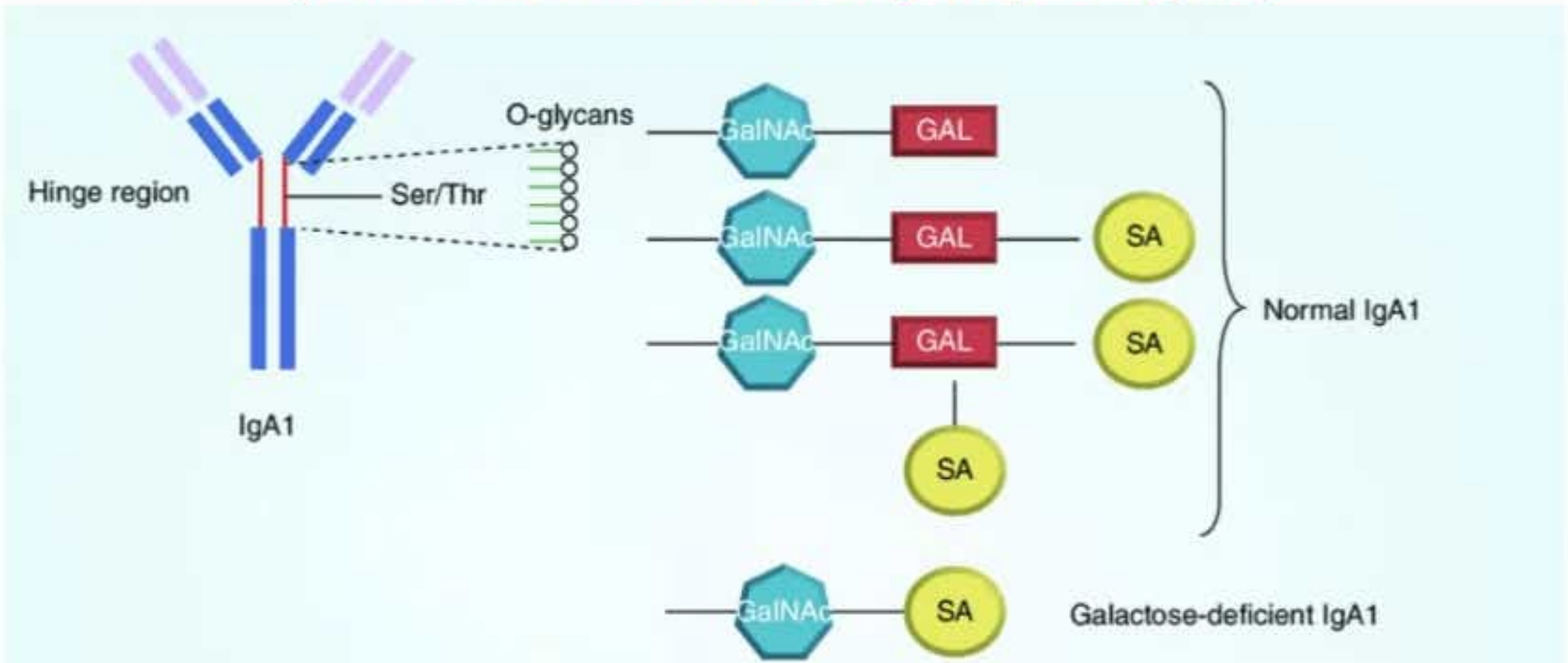
(Foot processes)

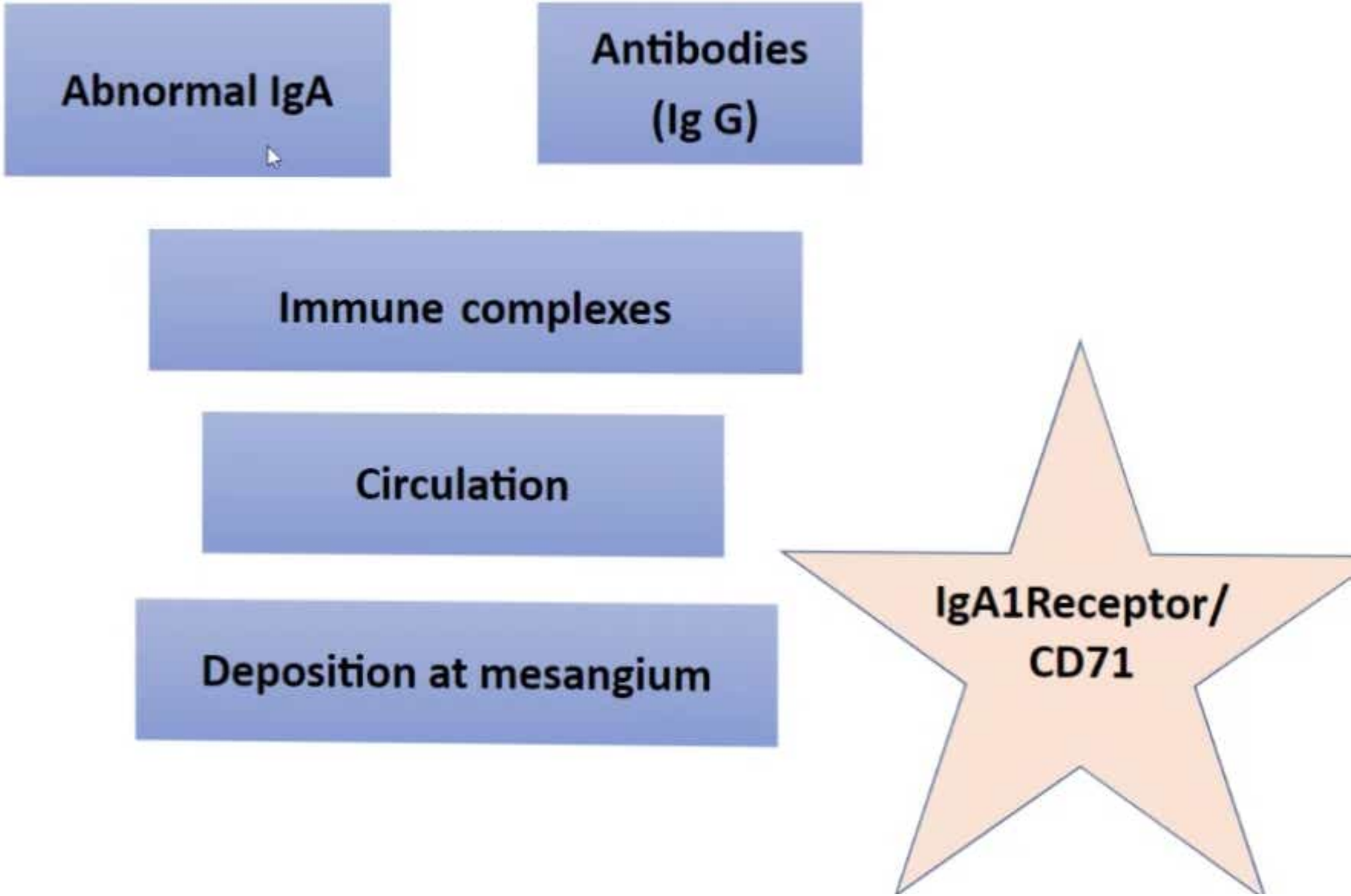
(GBM)

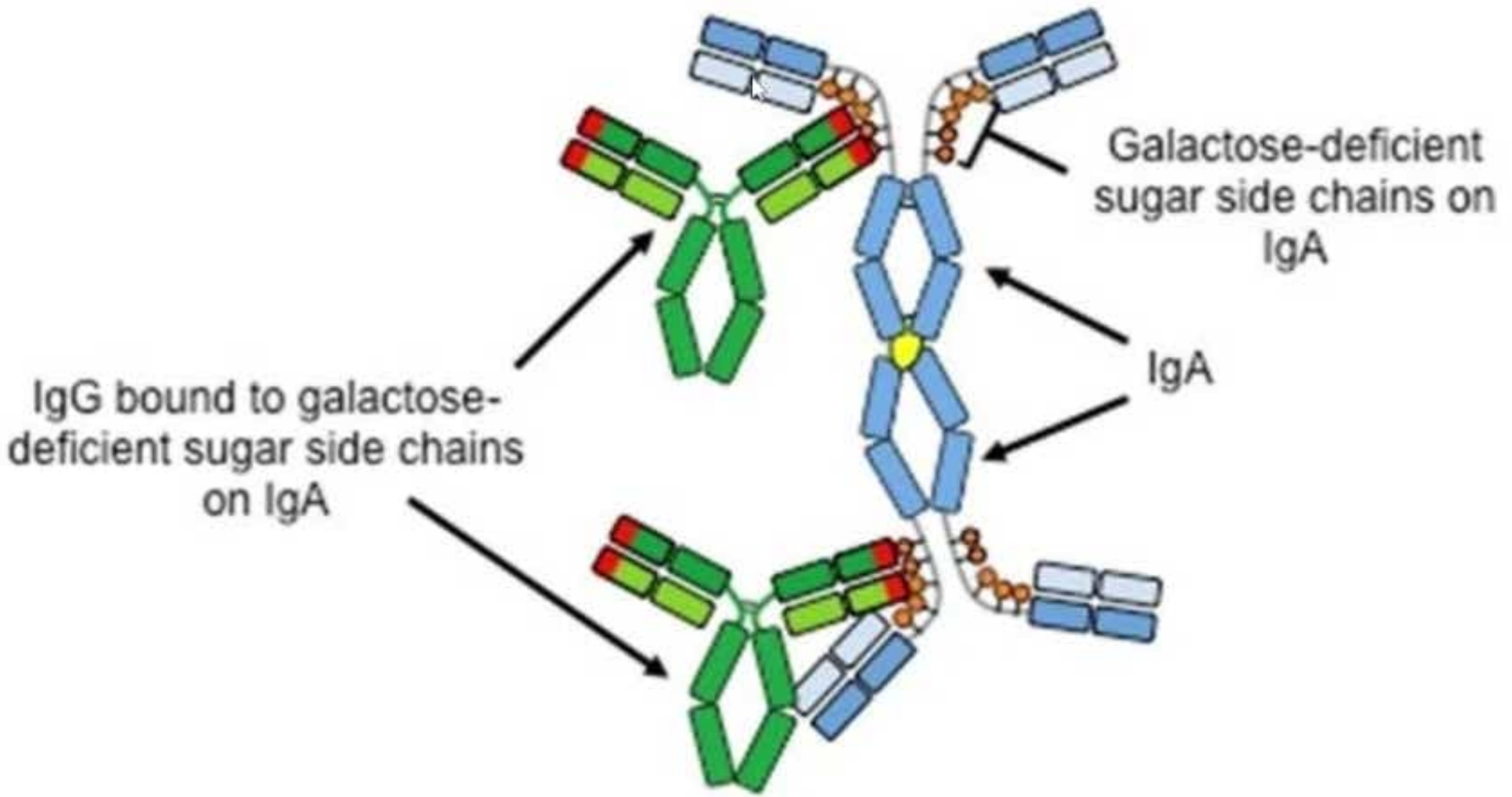
Pathogenesis

Abnormally glycosylated IgA1

(Galactose deficient Ig A1/ GdIgA1)




Abnormal IgA
Antibodies
(Ig G)
Immune complexes
Circulation
Deposition at mesangium
IgA1Receptor/
CD71


Galactose-deficient sugar side chains on IgA
IgA
IgG bound to galactose-deficient sugar side chains on IgA

Activation of Alternate Complement Pathway

↓

Production of proinflammatory cytokines

↓

Glomerular injury

↓

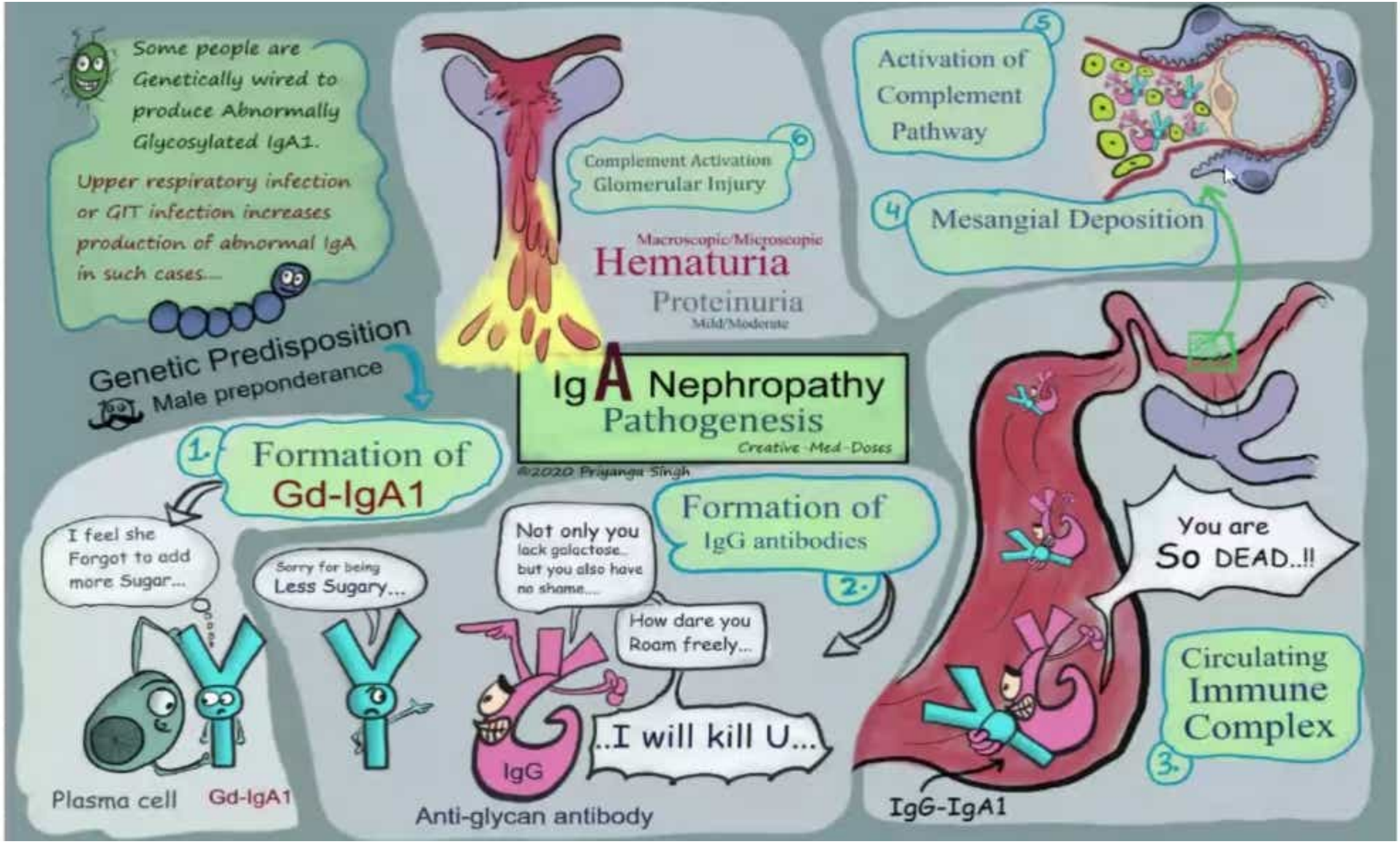
Filtration defect

↓

RBCs in urine

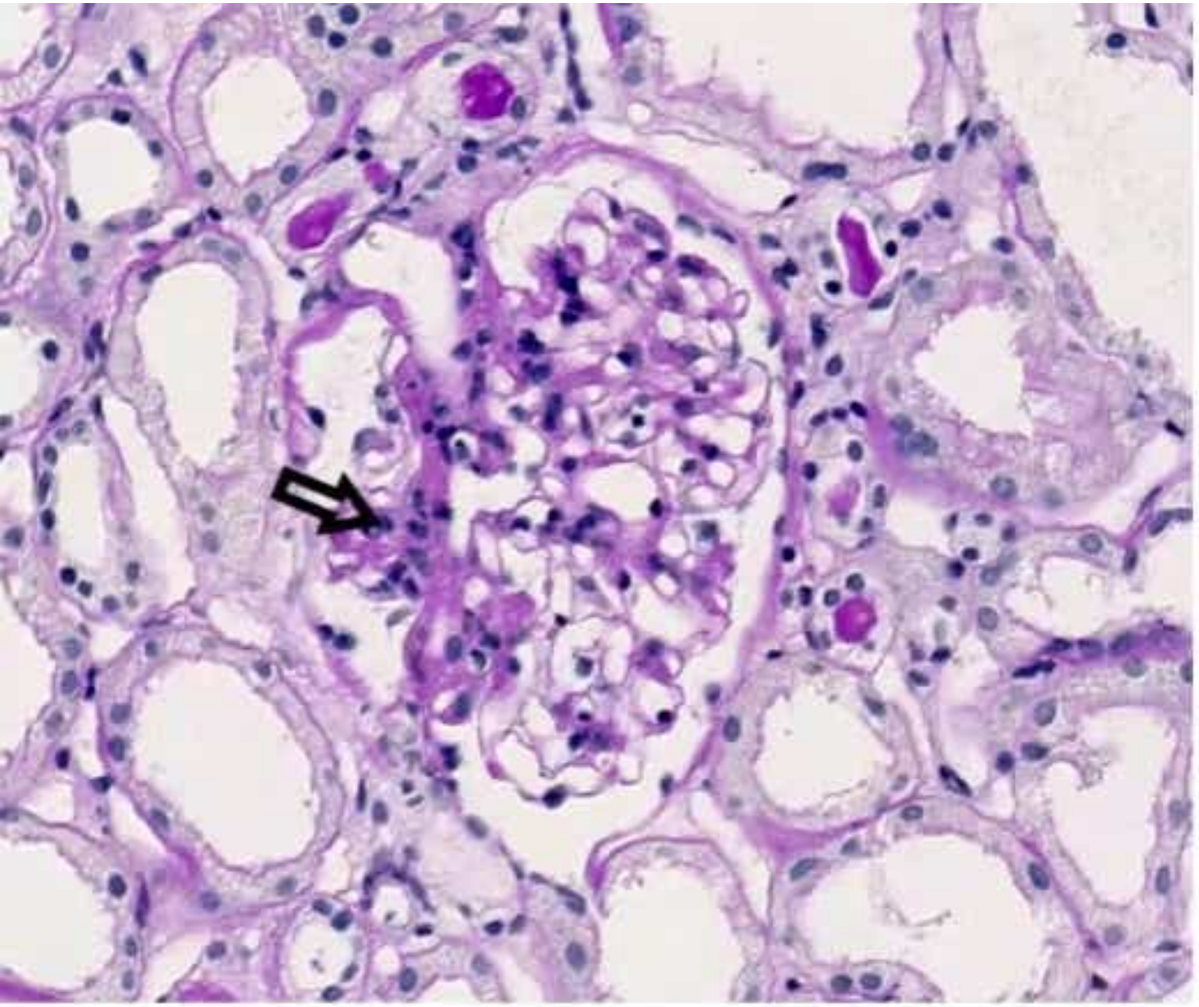
↓

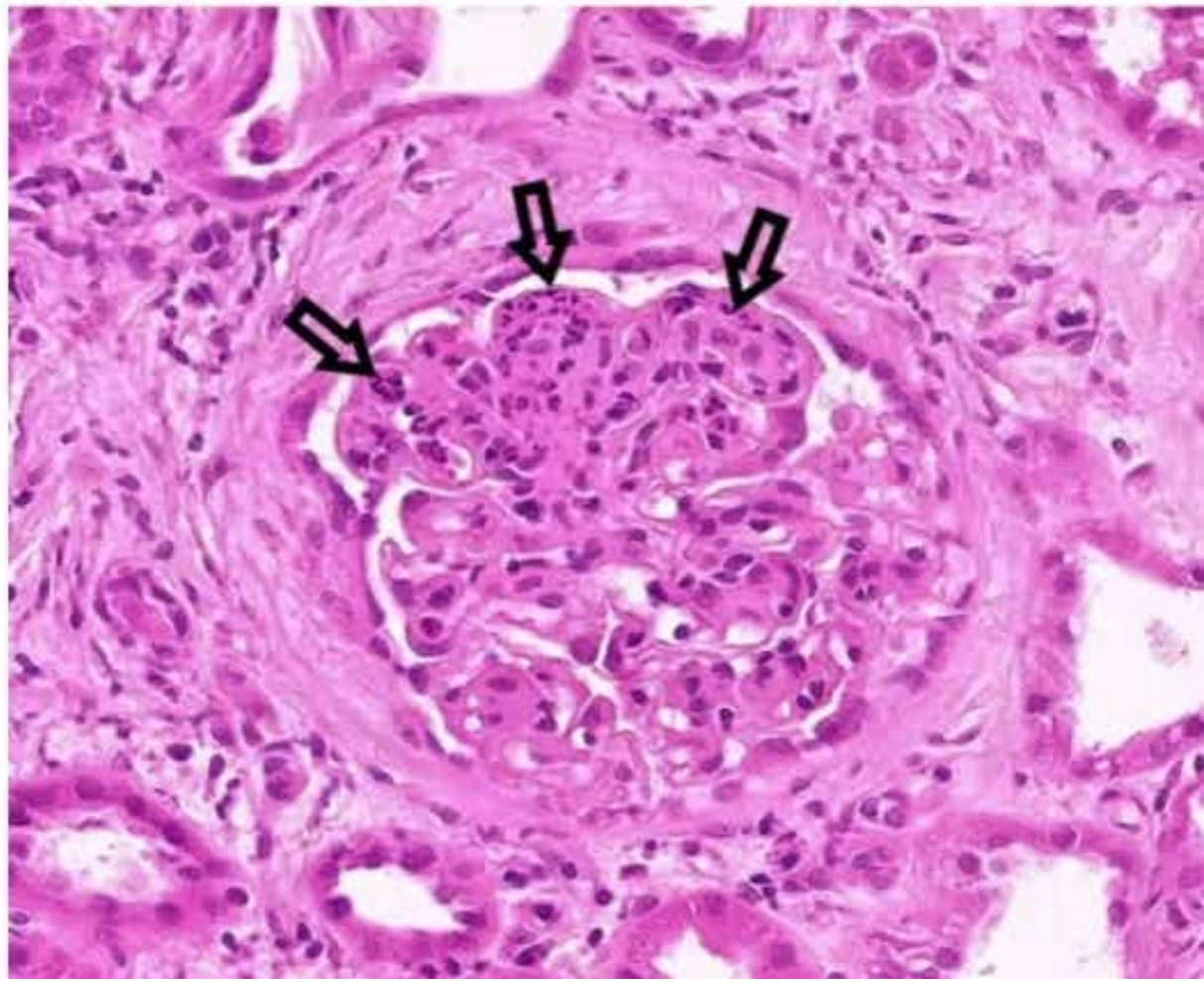
Hematuria

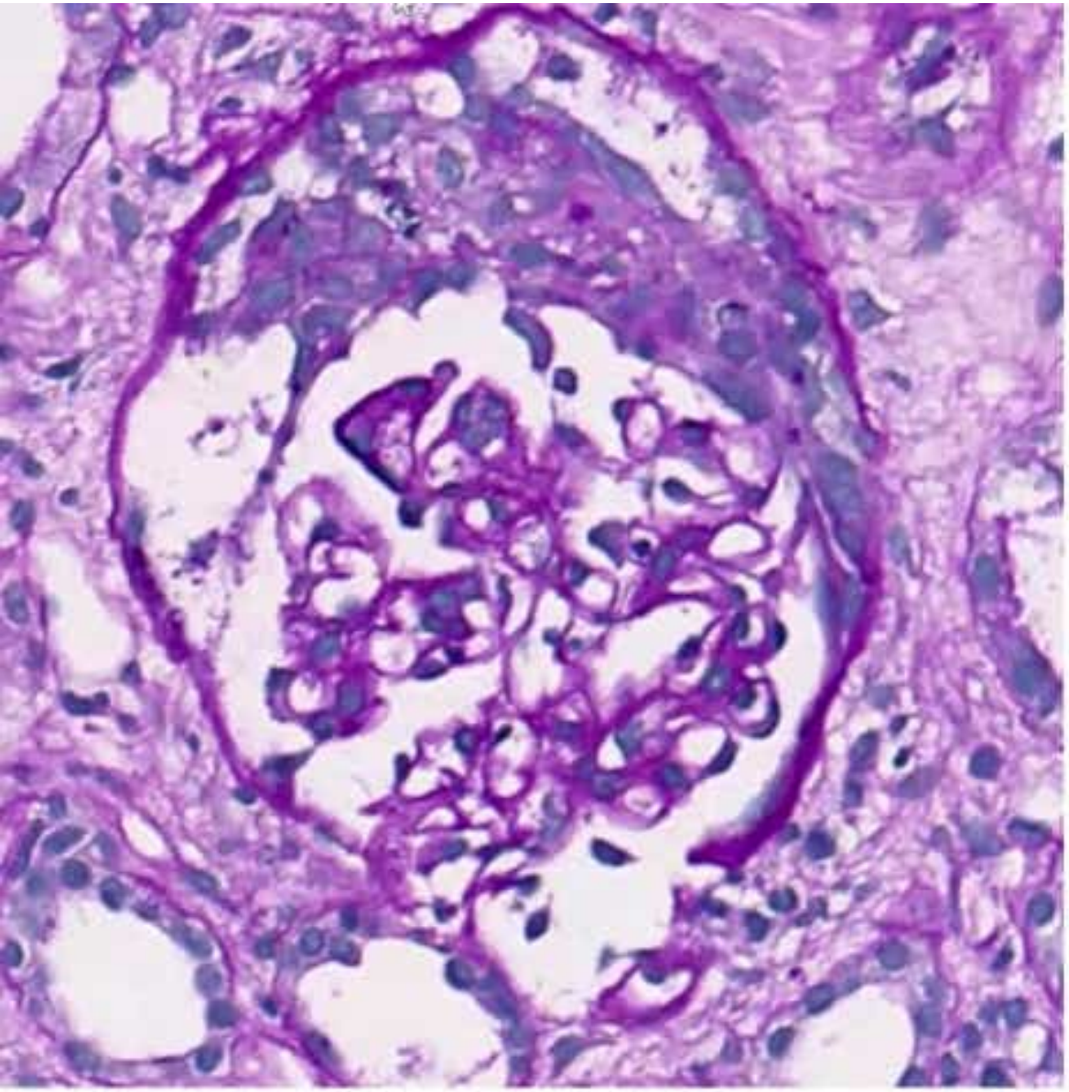

Some people are Genetically wired to produce Abnormally Glycosylated IgA1.
Upper respiratory infection or GIT infection increases production of abnormal IgA in such cases...
Genetic Predisposition
Male preponderance
Macroscopic/Microscopic
Hematuria
Proteinuria
Mild/Moderate
6
Complement Activation
Glomerular Injury
5
Activation of Complement Pathway
4
Mesangial Deposition
IgA Nephropathy
Pathogenesis
Creative-Med-Doses
©2020 Priyanga Singh
1.
Formation of
Gd-IgA1
I feel she Forgot to add more Sugar...
Sorry for being Less Sugary...
Plasma cell
Gd-IgA1
Formation of
IgG antibodies
2.
Not only you lack galactose.. but you also have no shame....
How dare you Roam freely...
..I will kill U...
IgG
Anti-glycan antibody
You are So DEAD..!!
Circulating
Immune
Complex
3.
IgG-IgA1

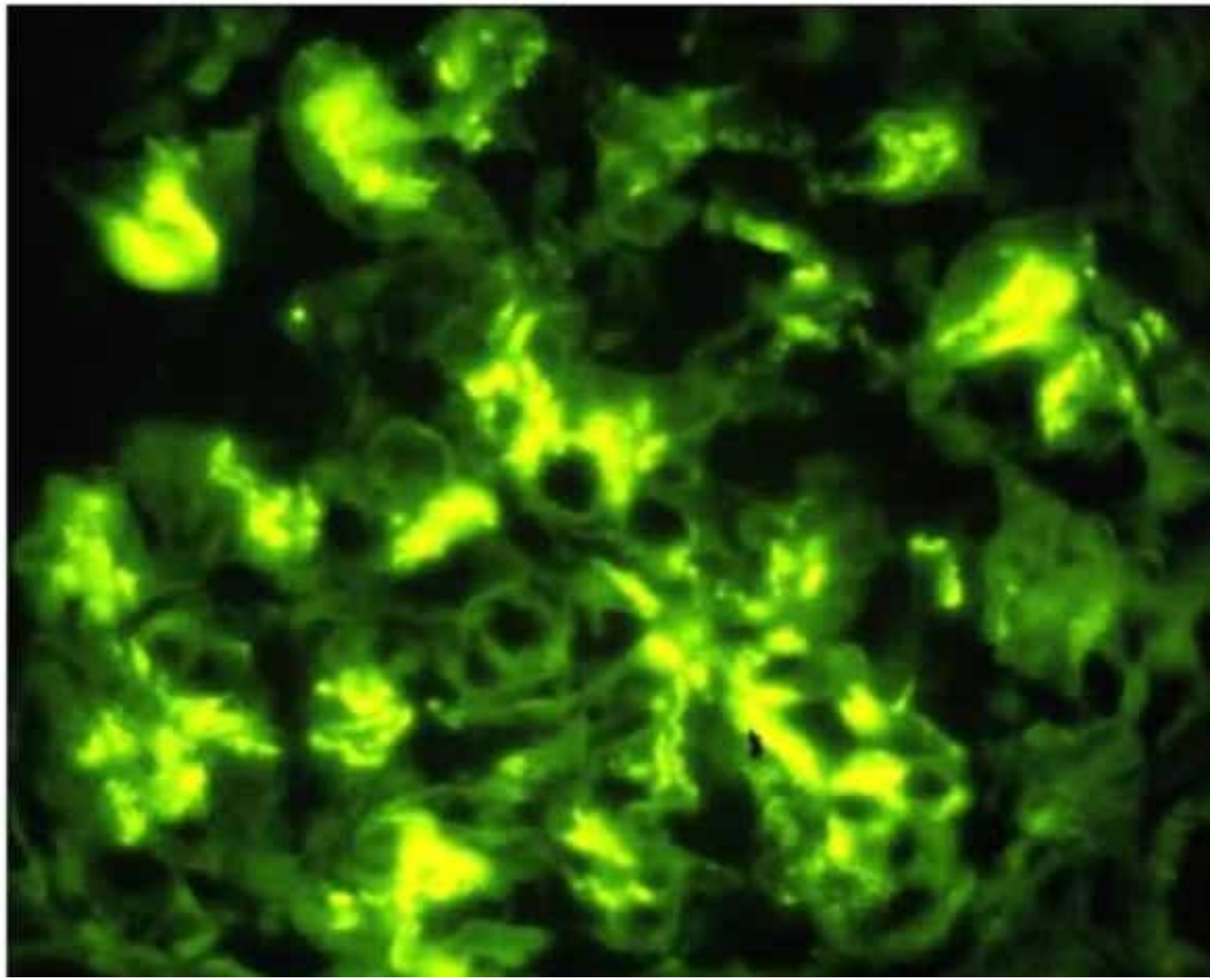
Morphology

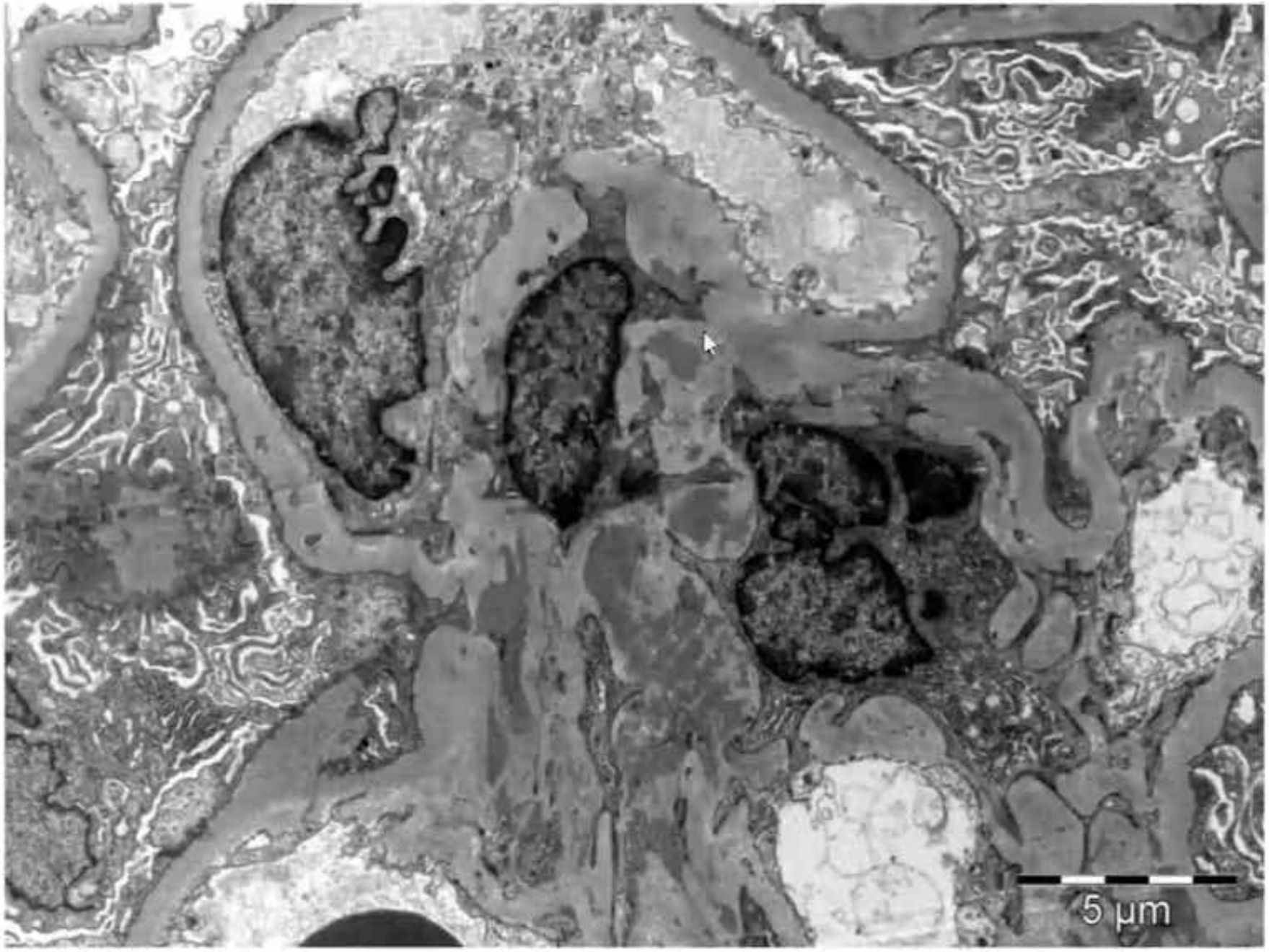
Light Microscopy	Immunofluorescence	Electron Microscopy
Normal	**Mesangial deposition of IgA**	**Electron dense deposit in mesangium**
Focal proliferative Glomerulonephritis (Mesangial widening/expansion/proliferation + segmental inflammation)		
Diffuse mesangial proliferation (Mesangioproliferative Glomerulonephritis)		
Overt Crescenteric Glomerulonephritis (rarely)		










5 µm

Clinical features

- Children & yound adults
- Gross hematuria
- Microscopic hematuria
- With or without proteinuria

- **Nephritic Syndrome**

- Following an episode of URTI/GIT/UTI

- Lasts for several days----subsides
  ----periodically occur

- Variable course

- For decades---normal----
  end stage renal disease over 20 years
  (20 – 50%)