NEFRITIK SINDROM

PROTEINURIYA + GEMATURIYA bilan kechadi.

Zararlanadi → GLOMERULYAR BAZAL MEMBRANA VA KAPILLIYAR ENDOTELIY

! Yallig'lanish bo'ladi.

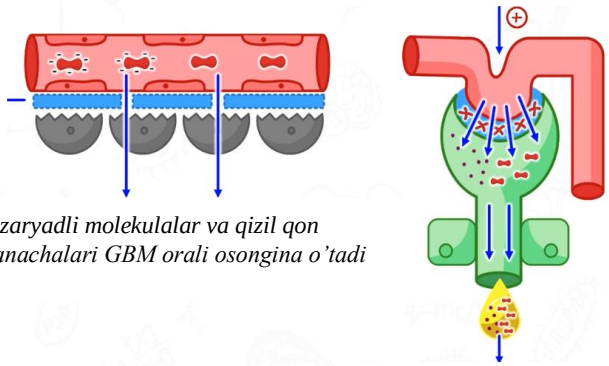
→ Endoteliyning o'lchamli barrieri

→ GBM ning zaryadli barrieri buzuladi

→ Shuning uchun asosan siydik orqali oqsil va qon chiqariladi.

GBMning zararlanishi va yallig'lanishi

GBMning zararlanishi



- zaryadli molekulalar va qizil qon tanachalari GBM orali osongina o'tadi

KLINIK BELGILARI:

- Shish: oyoqlarda, qovoqda, assit
- Gematuriya
- Proteinuriya <3,5g/kuniga
- Charchoq (azotemiya)
- Xolesterol miqdori ortishi

ASOSIY TURLARI:

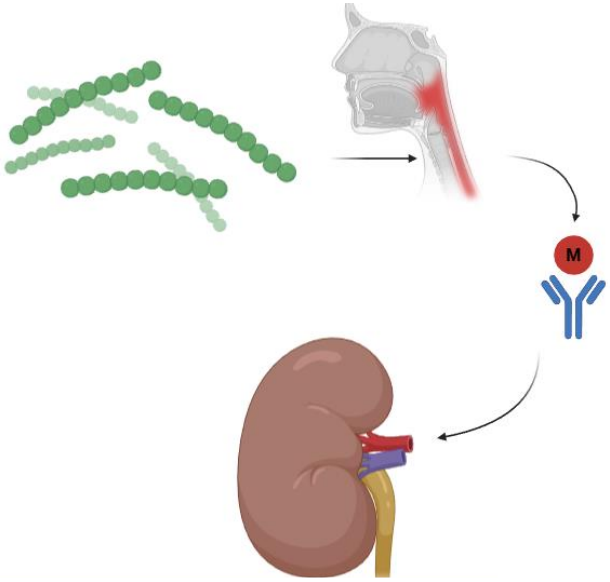
- Poststreptokokkal glomerulonefrit
- Berger (IgA) nefropatiyasi
- Diffuz proliferativ glomerulonefrit
- Tez rivojlanuvchi glomerulonefrit
- Alport sindromi
- Membranoproliferativ glomerulonefrit

POSTSTREP. GLOMERULONEFRIT

→ A guruhdagi β gemolitik streptokokkli infeksiyalar (impetigo, faringit) → M protein (nefritogenik)

→ Immun kompkeslar hosil bo'ladi va buyrakda depozitsiyalanadi.

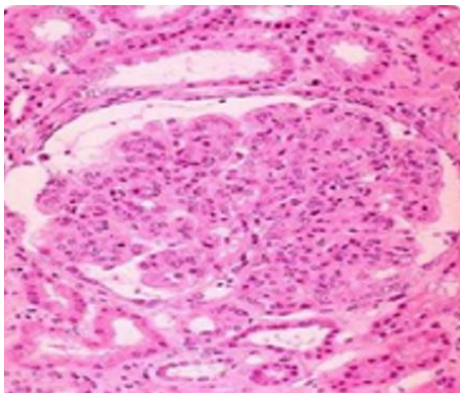
→ bu immun komplekslar komplemet oqsillarni biriktiradi va yallig'lanish hujyaralari (polimorfonuklear hujayralar) ni chorlaydi.



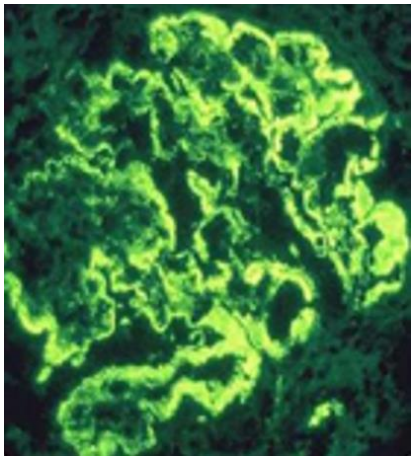
Bolalarda faringitdan so'ng yuzaga keladi.
Serologik testda → streptokokk
Ko'pincha o'z –o'zidan tuzalib ketadi

Kattalarda → yuqori nafas yo'llari infeksiyasi tufayli emas
Serologik test qilib bo'lmaydi → kultura olinadi
Buyrak yetishmovchiliggacha rivojlanishi mumkin.

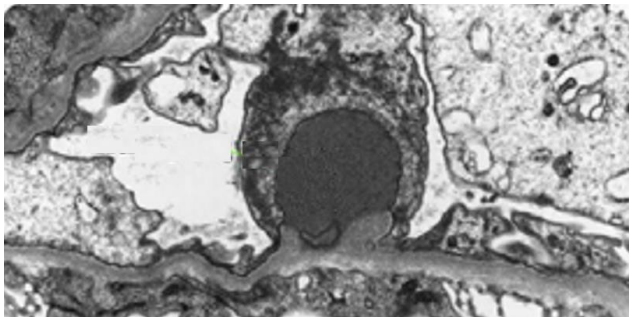
Yorug'lik mikroskopiyasida: gipersellular va kattalashgan glomerulus



- **Immunofluoressensiyada:** subendotelial IgG IgM va C_3 depozitsiyalar → granulyar ko'rinish



- **Elektron mikroskopiyada:** Subepitelial immunkompleks depozitsiyasi



bu kasallikda maxsus davo tadbirlari yo'q, ko'pincha o'z –o'zidan sog'ayib ketadi.

IgA NEFROPATIYASI

→ Glomerulonefritning dunyo bo'yicha eng ko'p tarqalgan shakli.

Qaytalanuvchi gematuriya epizodlari bilan kechadi.

- Nafas yo'llari infeksiyasi
- Oshqozon –ichak tizim infeksiyasi

Shilliq qavat → IgA → immun kompleks → mezangial hujayralarda depozitsiyalanadi

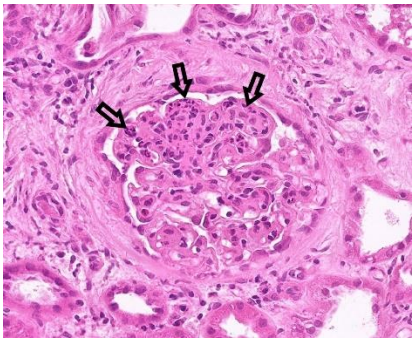
kompelent tizim aktivlanishi ←

↓

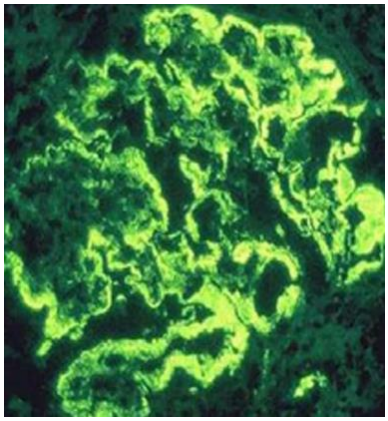
YALLIG'LANISH

! Poststrep. da gipokomplementemiya yuzaga keladi IgA nefropatiyada esa yo'q.

- **Yorug'lik mikroskopiyasida:** mezangial proliferatsiya



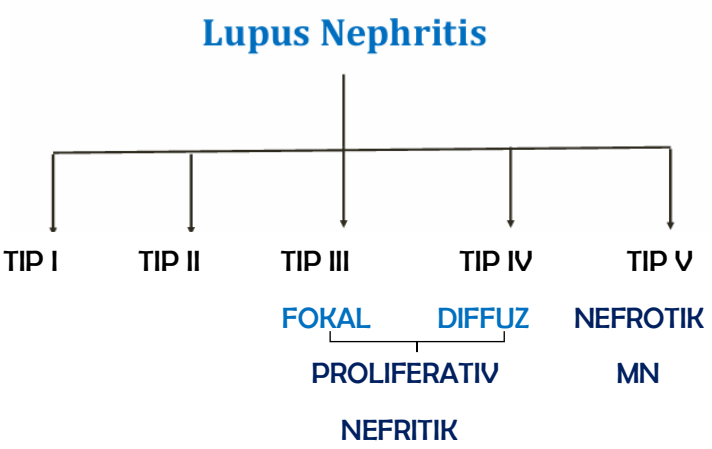
- **Immunofluoressensiyada:** mezangial granular depozitsiya (IgA) → elektron mikroskopiyada ham



Belgilar	PSGN	IgA nefropatiya
Infeksiya bilan bog'liqlik	1–3 hafta keyin	1–2 kun ichida
Yosh	Ko'proq bolalar	O'smir va yosh kattalar
Klinik boshlanish	O'tkir	Qaytalanuvchi
C3 komplement	↓ (pasaygan)	Normal
Streptokokk serologiya	ASO ↑, anti-DNaza B ↑	Normal
Light microscopy	Diffuz endokapillyar proliferatsiya, neutrofil ko'p	Mezangial proliferatsiya
Immunofluorescence	Granular "yulduzli osmon" (IgG + C3)	Mezangial granular IgA dominant
Electron microscopy	Subepithelial "bo'rtiqcha "lar	Mezangial depozitlar
Prognoz	Bolalarda yaxshi	Surunkalashishi mumkin

DIFFUZ PROLEFERATIV GLOMERULONEFRIT

Tizimli lupus eritramatoz → lupus nefrit IV



Diffuz → glomerulusning 50%dan ko'prog'i zarrlangan
Proliferativ → glomerulus hujayralari ortishi:

- mezangial
- endotelial
- monosit, neytrofil infltratsiyasi

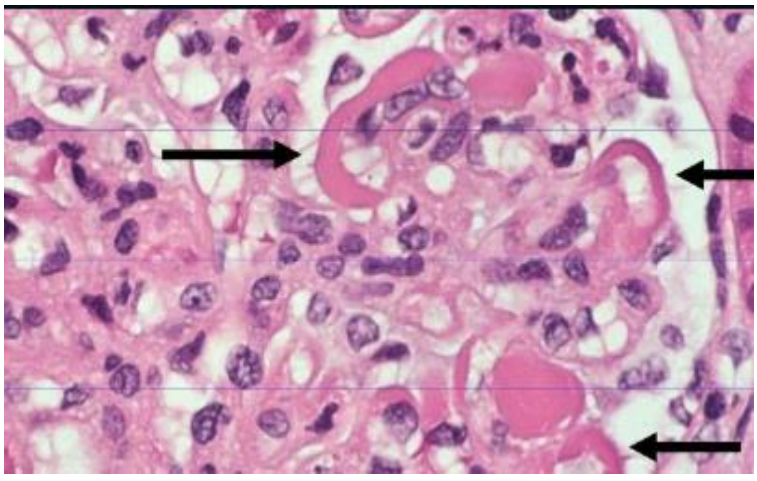
Ko'pincha lupus belgilari bilan birga kechadi:

- isitma
- qizil toshmalar
- artrit

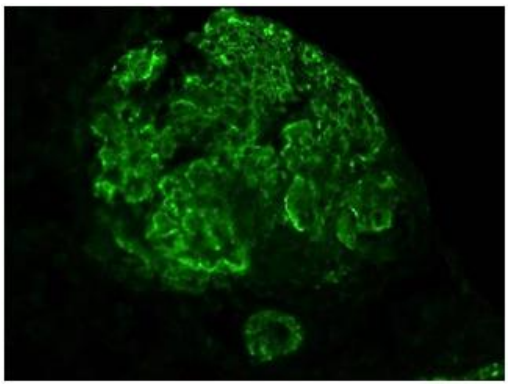
- Anti – ds DNA antitanalari → subendotelial depozitsiya

→ Komplement tizim aktivlashishi → autoimmun jarayon → glomerulonefrit (gipokomplementemiya)

- Yorug'lik mikroskopiyasida: simli halqa



- immunofluoressensiyada: granular



! Subendotelial, ba'zan subepitelial IgG, IgM, IgA, C_3 C_{1q} dan iborat depozitsiya.

→ Proteinuriya, gematuriya, filtratsiyaning pasayishi bilan namoyon bo'ladi.
→ Kopincha buyrak yetishmovchiligi rivojlanadi va gemodializ qilinadi.

ALPORT SINDROMI | X – D

IV tip kollagen α3 α4 α5 zanjirlar mutatsiyasi.

→ Glomerulyar bazal membrananing irregulyar yupqalashishi, qalinlashishi va ajralishi → gematuriya

Bu zanjirlar yana:
quloqda → sensorineural karlik
ko'zda → retinopatiya, anterior lenticonus

- Elektron mikroskopiyada: to'qilgan savat

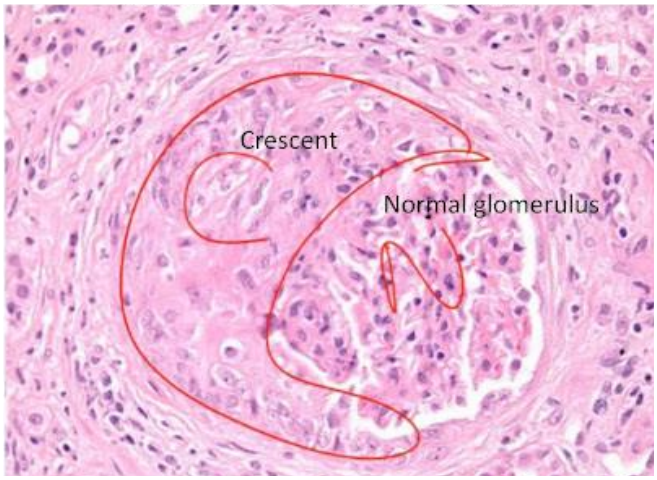
TEZ PROGRESSIV GLOMERULONEFRIT

- Glomerulonefritning juda og'ir formalaridan hisoblanadi

  → tezda o'tkir buyrak yetishmovchiligi rivojlanadi.

- KRESSENTIK (YARIMOYSIMON) glomerulonefrit ham deyiladi:

Yorug'lik mikroskopiyasida ko'ringanda



Yallig'lanish tufayli:

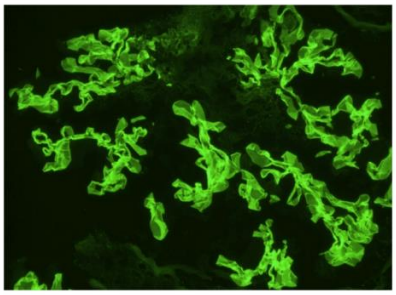
- yig'ilgan fibrin tolalar
- monosit
- makrofaglar
- glomerulyar pariteal hujayralarning haddan ortiq proliferatsiyasi

→ yarimoysimon shaklni hosil qiladi.

- Juda ko'p kasalliklar tez progressiyalanuvchi glomerulonefritga sabab bo'ladi. Bu kasalliklarning mexanizmini ajratib olsak bo'ladi:

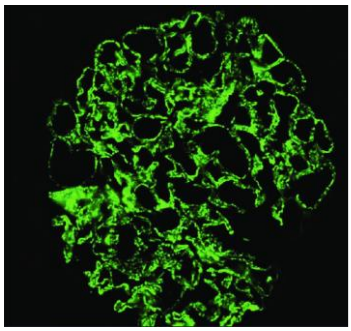
Immunofluressensiya usuli

LINEAR IF VIZUALIZATSIYA | TIP I



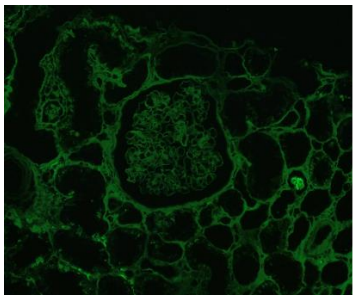
- "Goodpasture" sindromi
- Anti GBM antitana

GRANULAR IF VIZUALIZATSIYA | TIP II



- Poststreptokkokkal glomerulonefrit
- Diffuz proliferativ glomerulonefrit (Lupus)

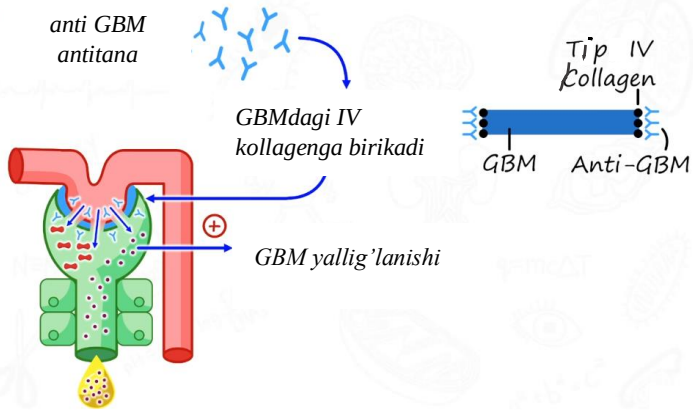
NEGATIV (PAUCI-IMMUNE) IF VIZUALIZATSIYA | TIP II

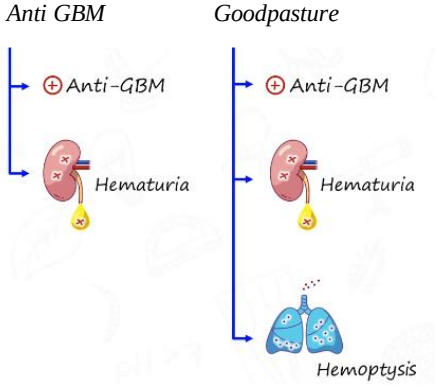


- Granulomatoz polyangit
- Mikroskopik polyangit
- Eozinofilik granulomatoz polyangit (Lupus)

GOODPASTURE VA ANTI GBM SINDROM

→ 2-tip gipersezuvchanlik (IgG)
→ Glomerular bazal membranadagi IV kollagernning α3 zanziriga qarshi





ANCA VASKULITLAR

→ Anti Neutrophil Cytoplasmic Antibodies

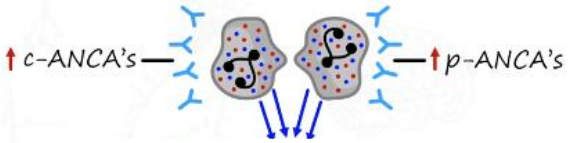
→ Ig va C_3 depozitlari bo'lmaydi.

p – ANCA:

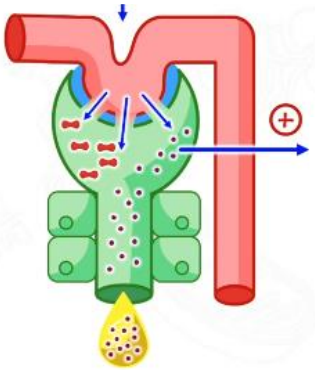
- mikroskopik polyangit
- eozinofilik granulomatoz polyangit

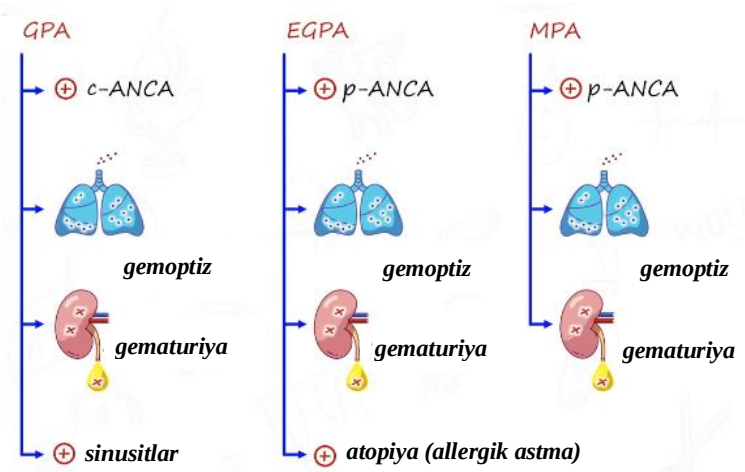
c – ANCA:

- granulamatoz polyangit



Kislorod radikallari va proteazalar





MEMBRANOPROLIFERATIV GLOMERULONEFRIT

NEFRITIK + NEFROTIK SINDROM

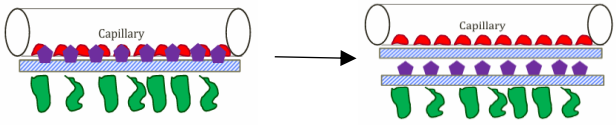
→ Kam uchraydi, proteinuriya (+/- nefrotik), gematuriya va buyrak yetishmovchiligi bilan kechadi.

Membrano → bazal membrana qalinlashishi

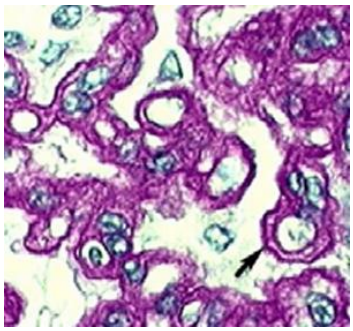
Proliferativ → mezangial hujayralar va matriksning proliferatsiyasi.

➢ TIP I → idiopatik yoki gepatit B/C bilan bog'liq bo'lishi mumkin

- Subendotelial immun kompleks (IgG) depozitsiyasi
- Bu depozitlar bazal membranani ikkiga ajratishi mumkin:



Natijada yorug'lik mikroskopiyasida huddi tramvay yo'lidek ko'rinish yuzaga keladi.

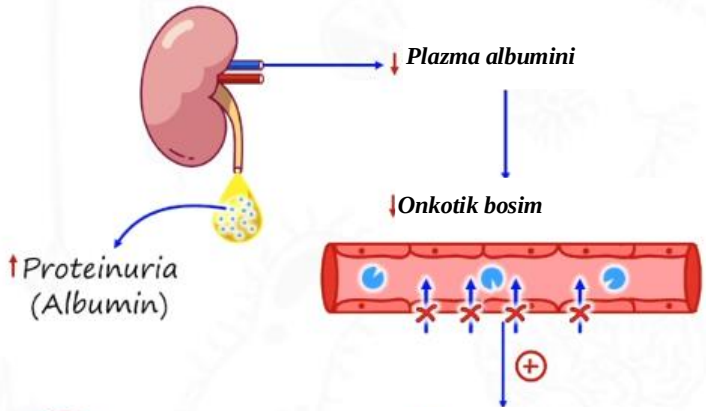


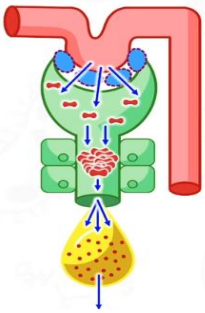
Immunofluoressensiyada → IgG va C_3 depozit (granular)

➢ TIP II → C_3 konvertaza stabillovchi antitana

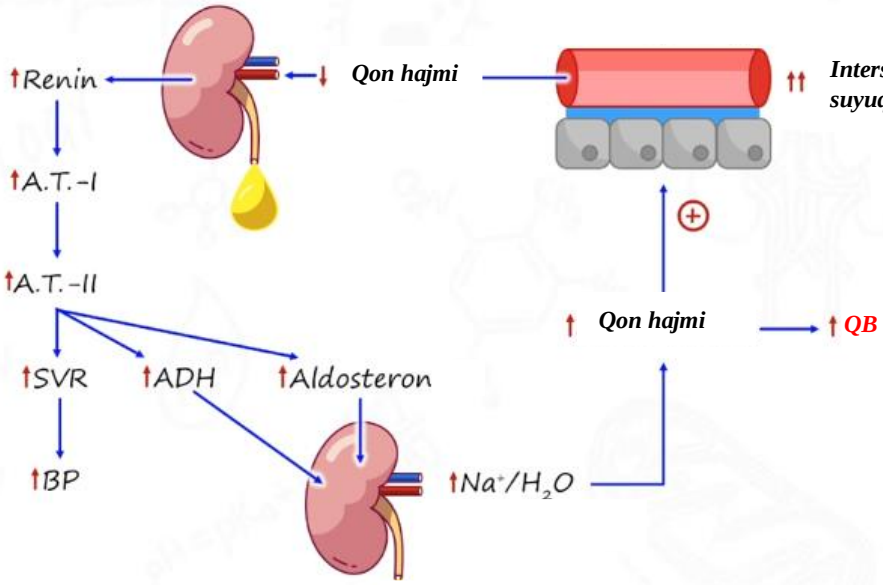
- C_3 aktivlanib ketishi → intramembranoz C_3 depozit.
- Bu holatda ham yorug'lik mikroskopiyasida → tramvay yo'li ko'rinishi.

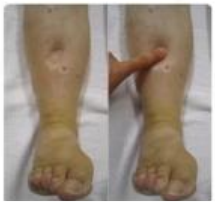
NEFRITIK SINDROM | ASORATLARI

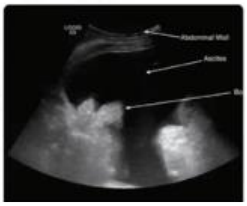


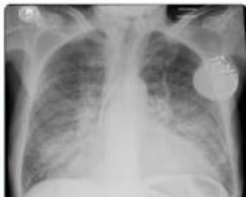


Gematuriya
Qizil qon hujayralari, akantositlar

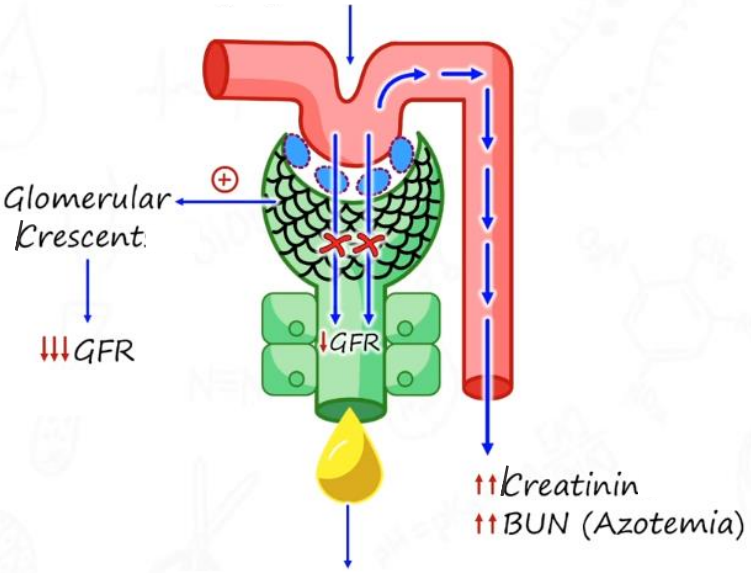




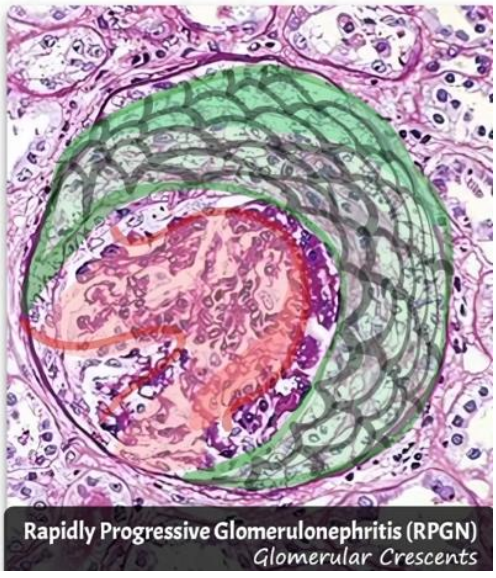


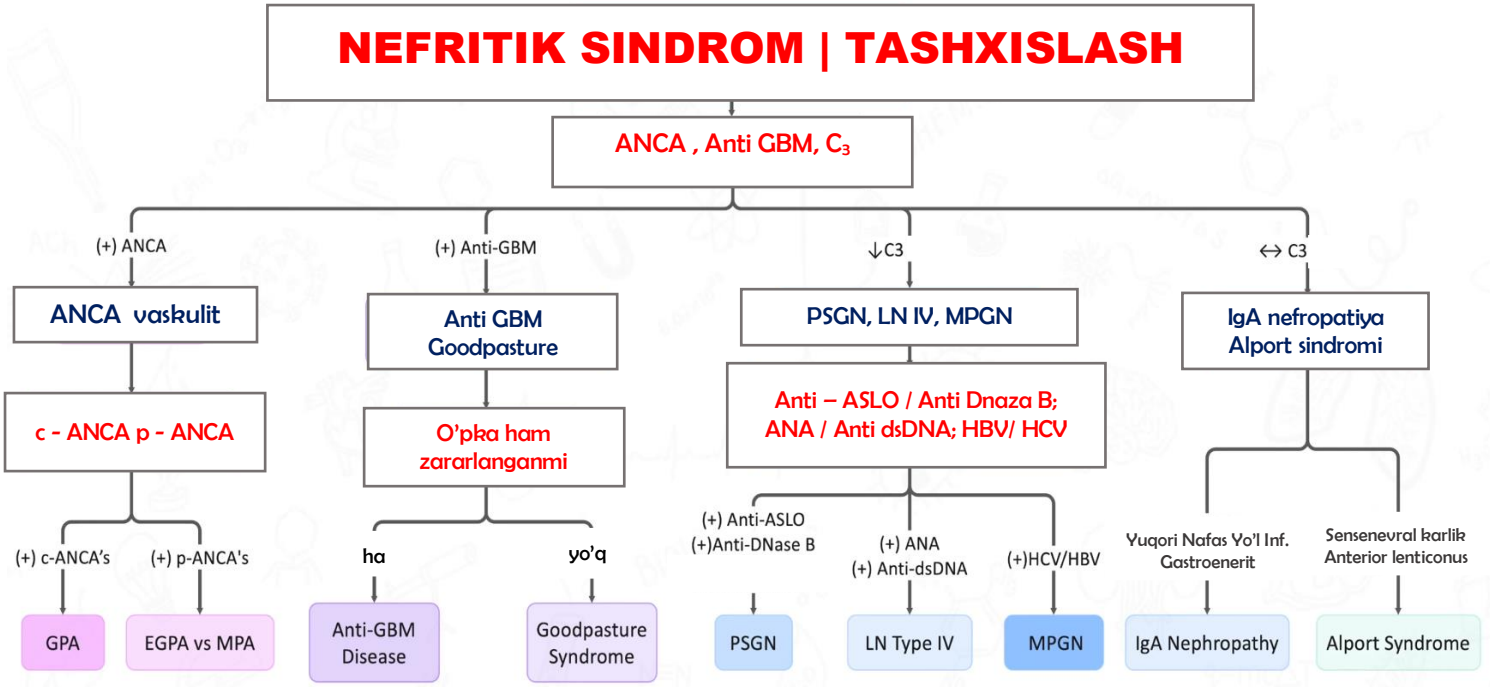


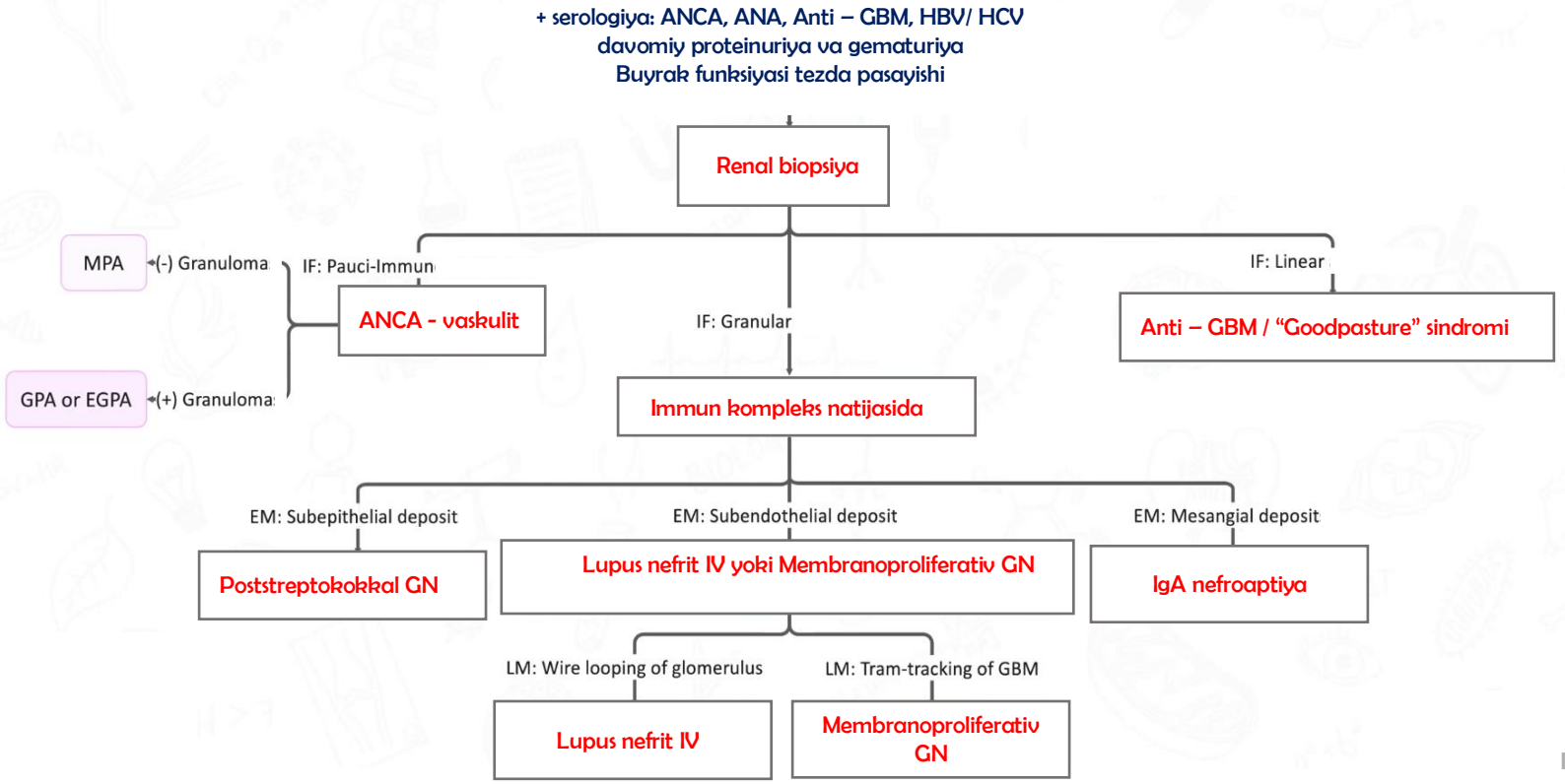
Glomerulyar zararlanish



Oligouriya

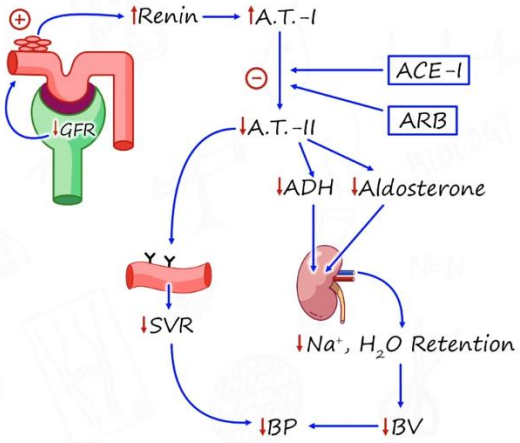


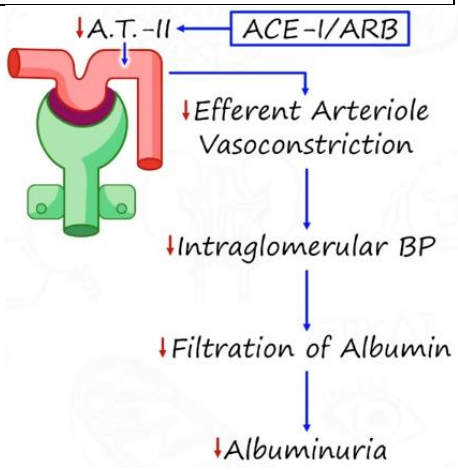

NEFRITIK SINDROM | TASHXISLASH
ANCA , Anti GBM, C3
(+) ANCA
(+) Anti-GBM
↓C3
↔ C3
ANCA vaskulit
Anti GBM Goodpasture
PSGN, LN IV, MPGN
IgA nefropatiya Alport sindromi
c - ANCA p - ANCA
O'pka ham zararlanganmi
Anti – ASLO / Anti Dnaza B; ANA / Anti dsDNA; HBV/ HCV
(+) c-ANCA's
(+) p-ANCA's
ha
yo'q
(+) Anti-ASLO (+)Anti-DNase B
(+) ANA (+) Anti-dsDNA
(+)HCV/HBV
Yuqori Nafas Yo'l Inf. Gastroenerit
Sensenevral karlik Anterior lenticonus
GPA
EGPA vs MPA
Anti-GBM Disease
Goodpasture Syndrome
PSGN
LN Type IV
MPGN
IgA Nephropathy
Alport Syndrome


+ serologiya: ANCA, ANA, Anti – GBM, HBV/ HCV
davomiy proteinuriya va gematuriya
Buyrak funksiyasi tezda pasayishi
Renal biopsiya
MPA
(-) Granuloma
IF: Pauci-Immun
IF: Linear
ANCA - vaskulit
IF: Granular
Anti – GBM / "Goodpasture" sindromi
GPA or EGPA
(+) Granuloma
Immun kompleks natijasida
EM: Subepithelial deposit
EM: Subendothelial deposit
EM: Mesangial deposit
Poststreptokokkal GN
Lupus nefrit IV yoki Membranoproliferativ GN
IgA nefroaptiya
LM: Wire looping of glomerulus
LM: Tram-tracking of GBM
Lupus nefrit IV
Membranoproliferativ GN

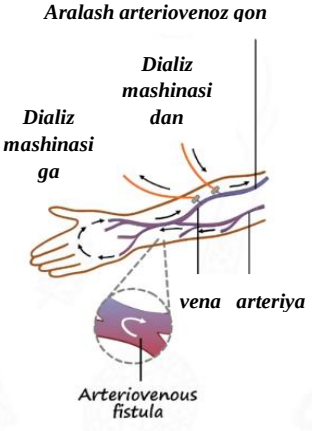
GLOMERULAR KASALLIKLAR | DAVOLASH

DAVOLASH USULLARI	KO'RSATMALAR
• Natriy iste'molini cheklash • Halqa diuretiklari	➢ Edema
• AKE ingibitorlari • Aldosteron retseptor blokatorlari	➢ Gipertenziya ➢ Proteinuriya
• Statin terapiya	➢ Giperlipidemiya
• Antikoagulyantlar	➢ Giperkoagulyatsiya
• Pneumoccocal vaksinatsiya	➢ Infeksiyaga moyillik ortganda
• Dializ	➢ Buyrak yetishmovchilik oxirgi bosqichlari

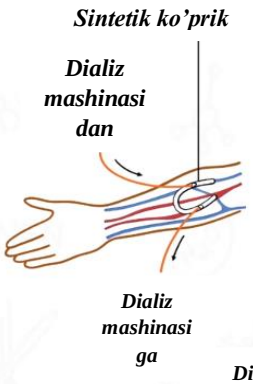




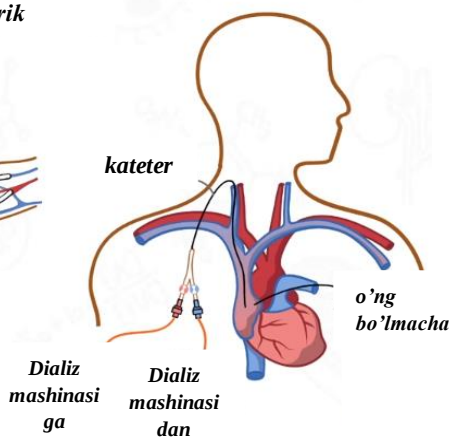
a) AV fistula



b) AV graft



c) Markaziy venoz kateter



DAVOLASH USULLARI	SINDROMGA SABAB BO'LGAN HOLATLAR
• Steroidlar	➢ Minimal o'zgarishli nefropatiya ➢ Iga nefropatiya
• Steroidlar + immunosupressantlar	➢ Fokal segmental glomuloskleroz ➢ Membranoz nefropatiya ➢ ANCA vaskulit ➢ Anti GBM kasallik ➢ Lupus nefrit IV tip ➢ Membranoproliferativ glomerulonefrit ➢ Poststrepptokokkal nefropatiya
• Plazmoferez	➢ Anti GMB kasallik ➢ ANCA vaskulit + alveolar gemorragiya ➢ Tez rivojlanuvchi glomerulonefrit