- N.V. 42 ani, etnie rroma
- 09.2014 – Landesklinikum Mistelbach-Ganserndorf - Viena
- CLINIC
  - Edeme generalizate
  - Crestere in greutate (10 kg/2 luni)
  - Astenie, fatigabilitate
  - BIOLOGIC

	Valori normale	19.09.2014	22.09.2014	30.09.2014
Creatinina serica	0,5 – 1 mg/dl	**1,8**	**1,7**	**1,2**
Uree serica	6 - 20 mg/dl	**42**	**27**	**24**
Proteine totale	6,4 - 8,3 g/dl		**4,6**	**5,8**
Albumine serice	3,5 - 5,2 g/l		**1,8**	**2,5**
Colesterol	<200 mg/dl			**450**
Trigliceride	30 – 150 mg/dl			**203**
TSH	0,44 - 3,77 µu/ml			**5,42**
FT3	0,76 - 1,66 ng/dl			**0,7**
Sumar de urina	Hematii (negativ) Proteine (mg/l) (negativ)		**0** **6000**	
Sediment urinar	Hematii		**0**	
Proteinurie	< 150 g/24 ore		**5,7**	

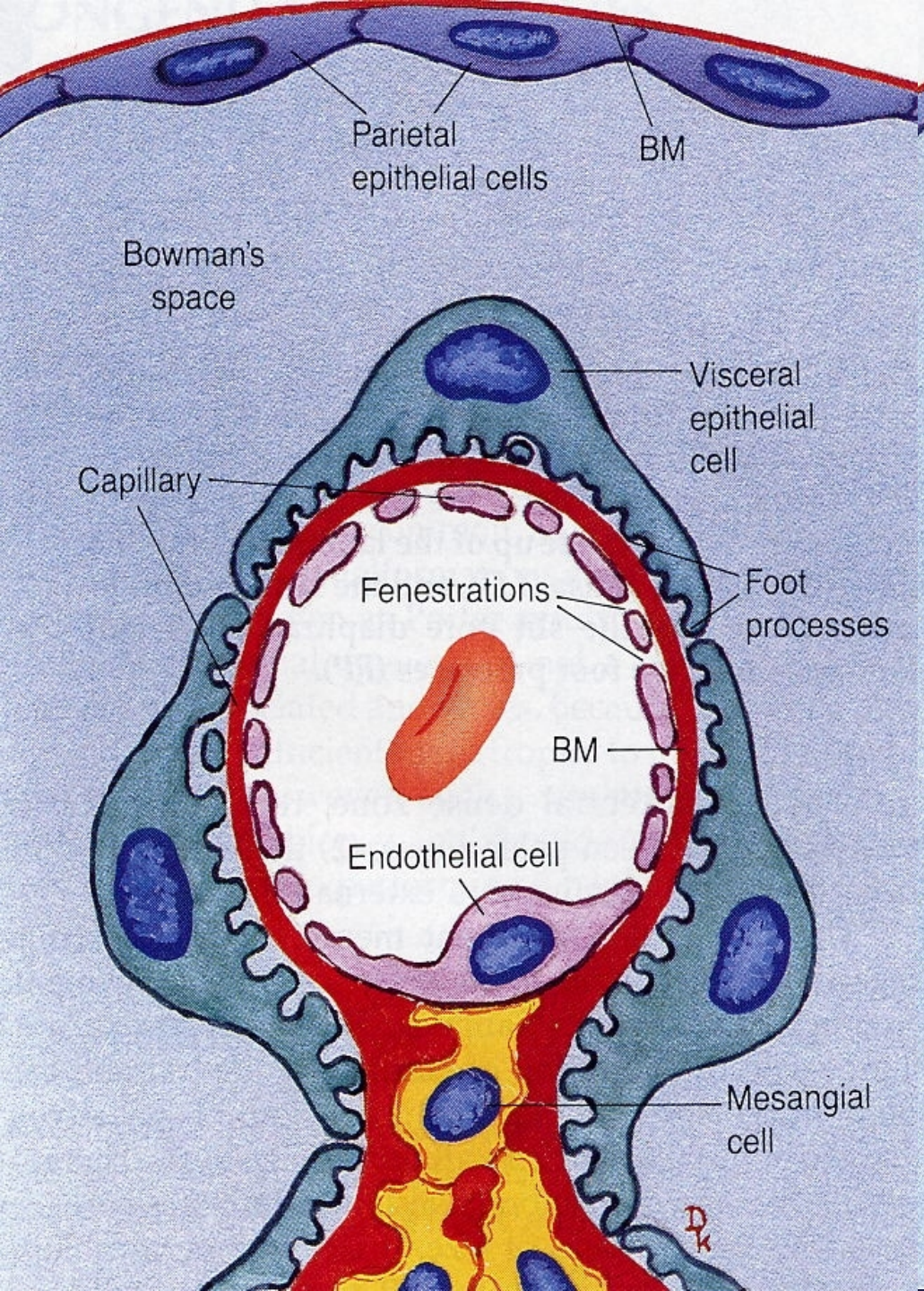
PUNCTIE BIOPSIE RENALA (17.09.2014)

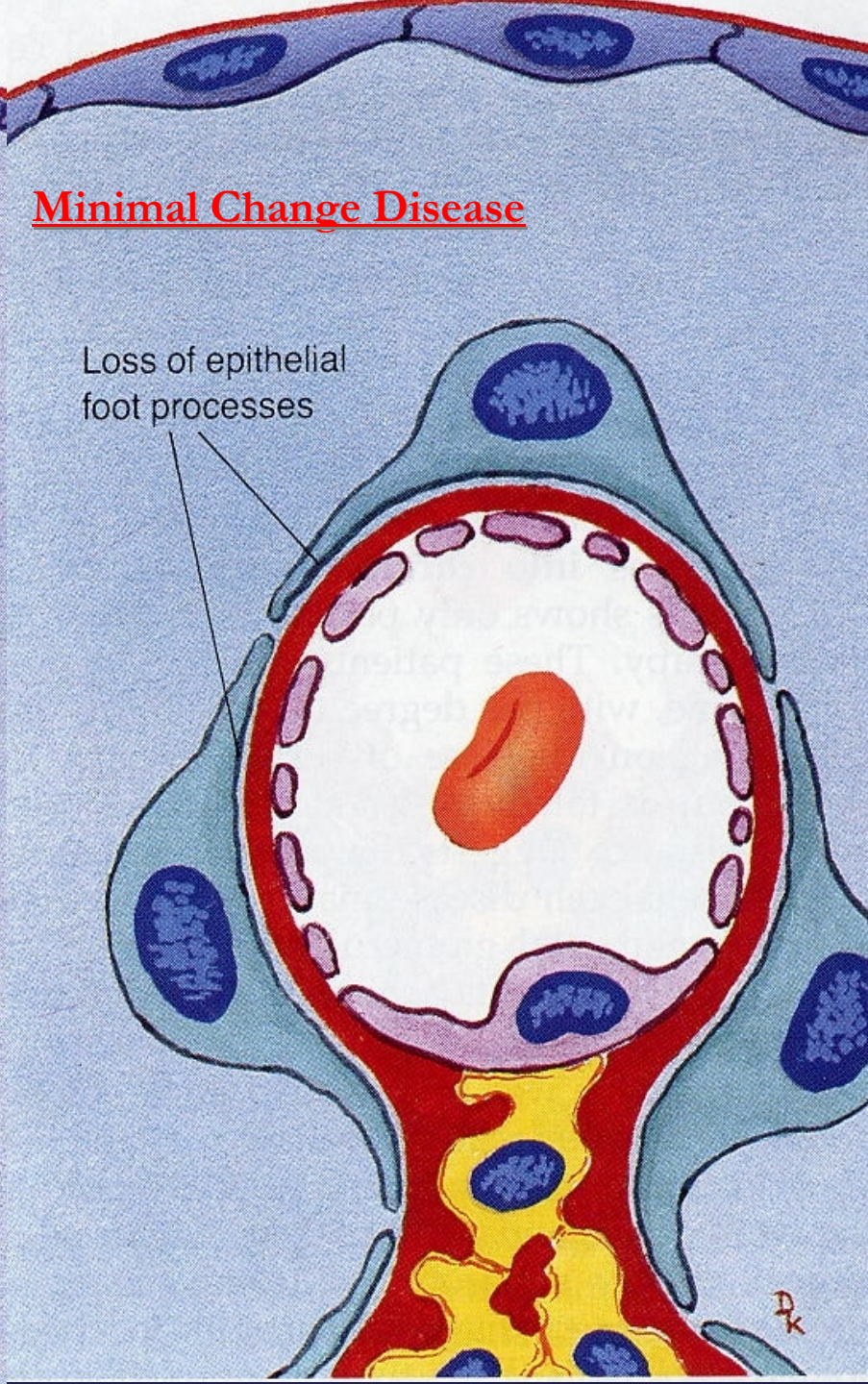
- 30 glomeruli
- MO (H.E, PAS, impregnare argentica): minima expansiune a matricei mezangiale

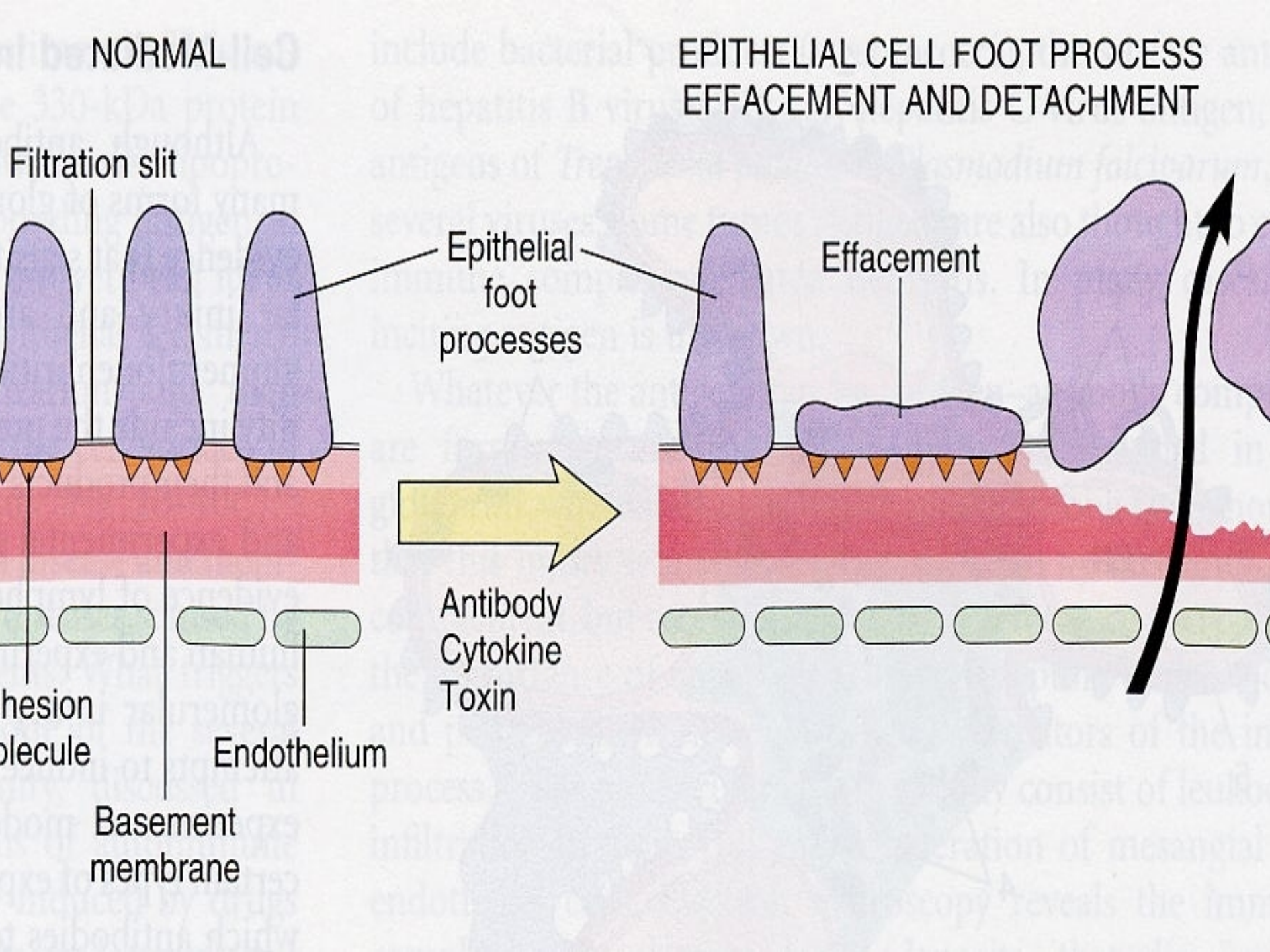
  fara: proliferare celulara, sinechii capsulare, semilune
- Fara leziuni tubulare sau interstitiale
- Fara modificari vasculare
- Imunohistochimie: minime depozite mezangiale de IgM, C1q si C3
- ME: stergerea difuza a proceselor podocitare

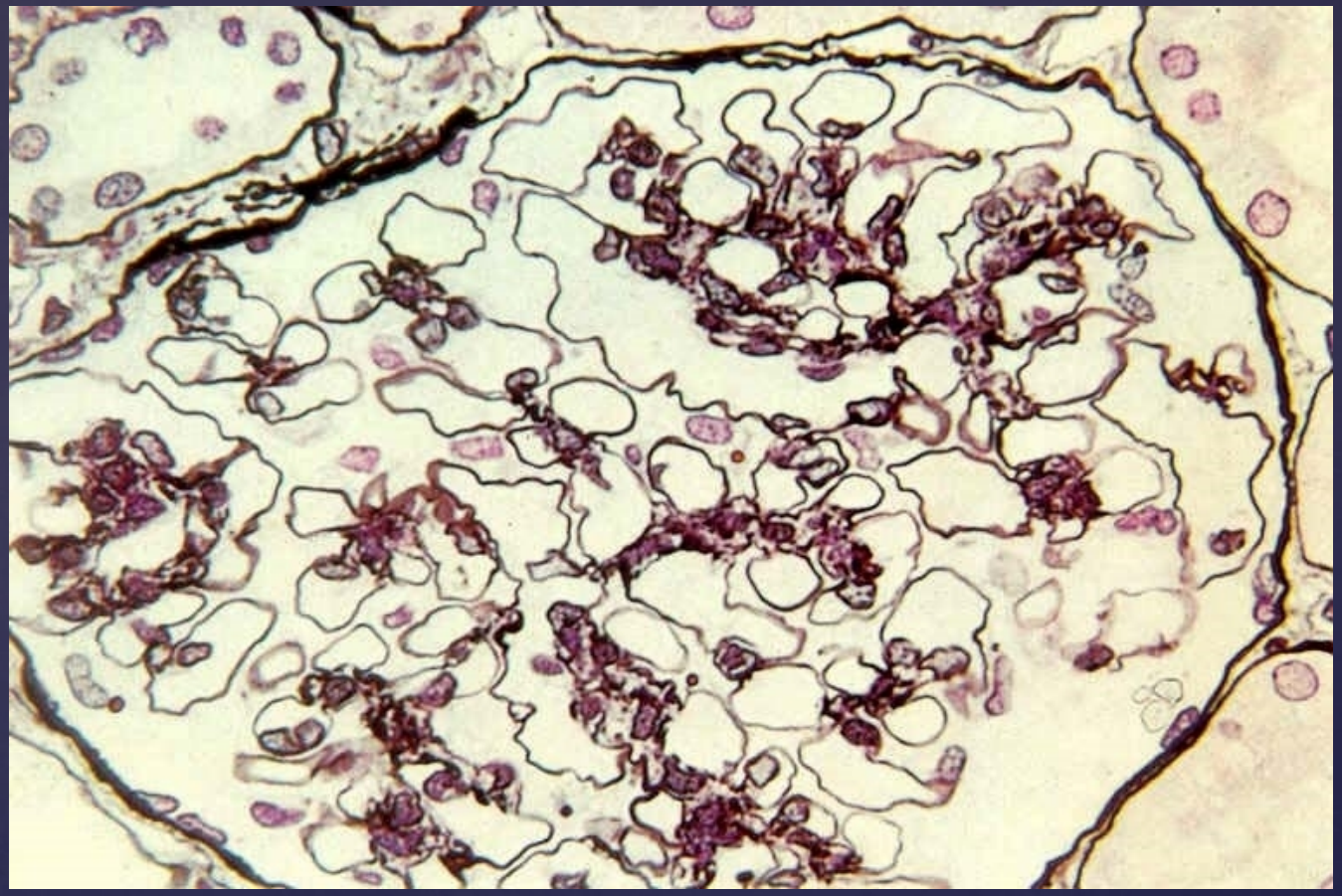
Concluzie: Nefropatie glomerulara cu leziuni minime

Minimal Change Disease

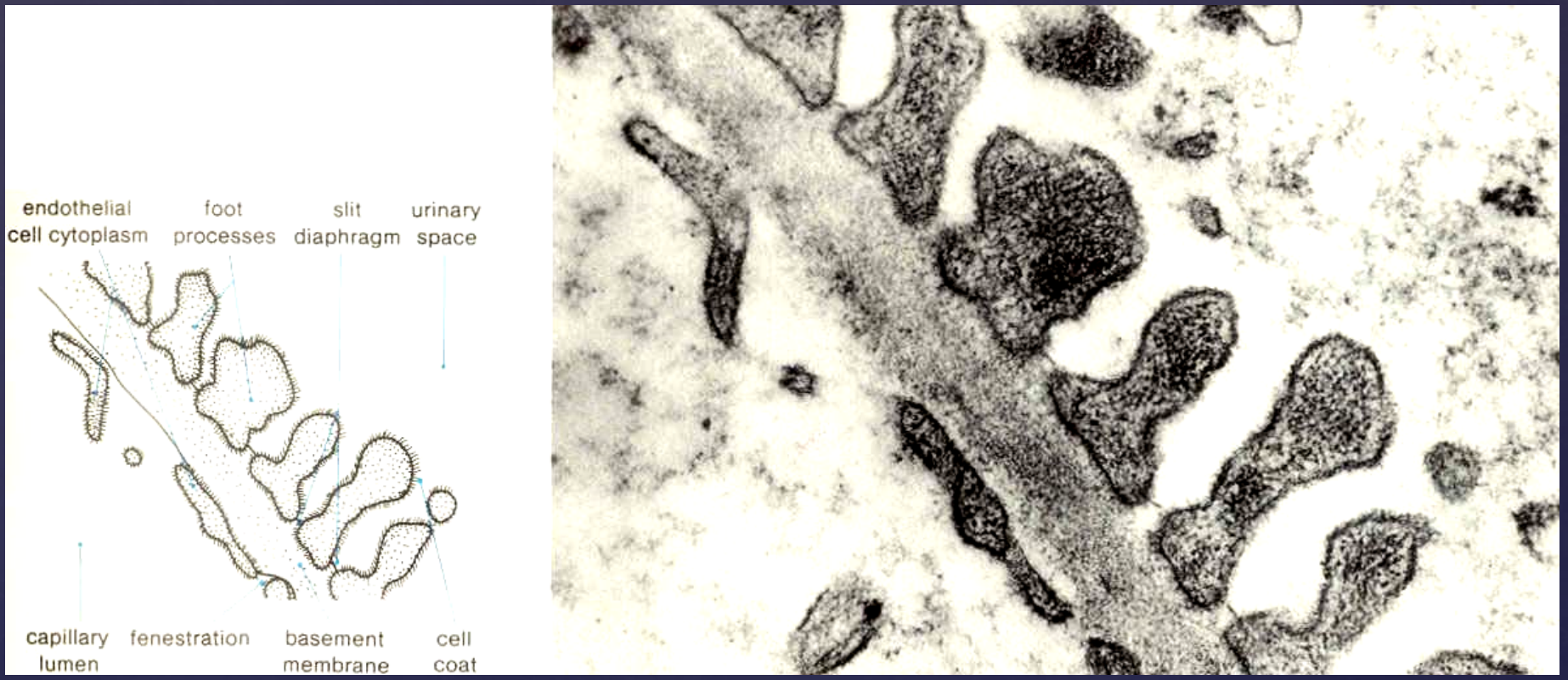





NORMAL
EPITHELIAL CELL FOOT PROCESS
EFFACEMENT AND DETACHMENT
Filtration slit
Epithelial
foot
processes
Effacement
Antibody
Cytokine
Toxin
hesion
olecule
Endothelium
Basement
membrane



[Nachman P.H., Jennette J.C., Falk R.J. Primary Glomerular Disease. Brenner & Rector's The Kidney 9th Edition, 2012]


endothelial cell cytoplasm
foot processes
slit diaphragm
urinary space
capillary lumen
fenestration
basement membrane
cell coat

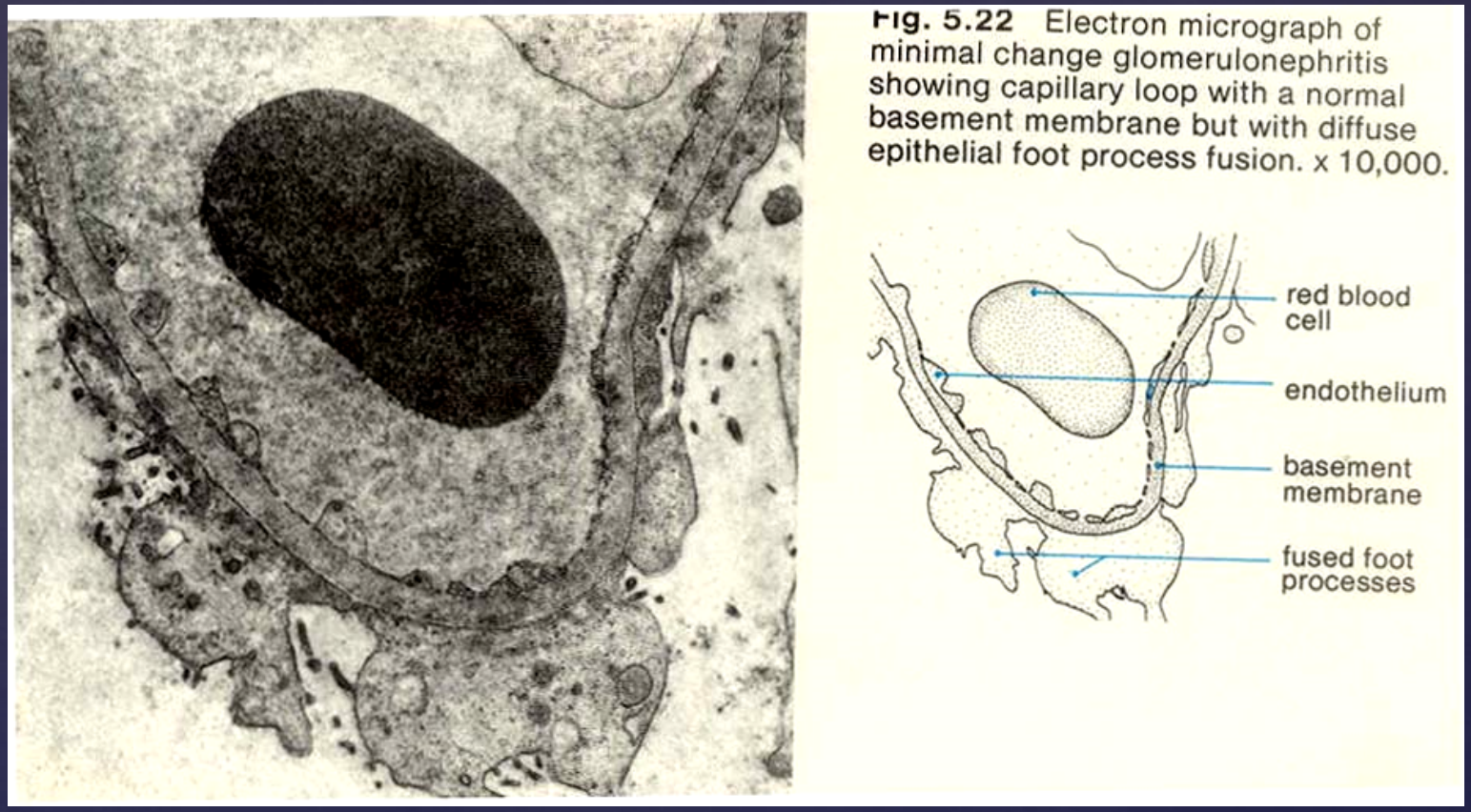


Fig. 5.22 Electron micrograph of minimal change glomerulonephritis showing capillary loop with a normal basement membrane but with diffuse epithelial foot process fusion. x 10,000.

Diagnostic pozitiv de etapa

- proteinurie 5,7 g/24 ore
- hipoalbuminemie 1,8 g/l
- sindrom edematos
- hiperlipemie

⇒ **SINDROM NEFROTIC**

Valori crescute ale retentiei azotate (creatinina serica = 1,8 mg/dl) cu normalizarea in 14 zile. Azotemia prerenala produsa prin hipoperfuzia renala in cadrul sindromului nefrotic, al hipoproteinemiei

=> LEZIUNE ACUTA DE RINICHI - AZOTEMIE PRERENALA

TA normala, sediment urinar fara modificari, biopsia renala cu glomeruli optic normali, cu fuziunea proceselor podocitare **=> NEFROPATIE CU LEZIUNI GLOMERULARE MINIME**

[Nachman P.H., Jennette J.C., Falk R.J. Primary Glomerular Disease. Brenner & Rector's The Kidney 9th Edition, 2012]

Diagnostic diferential

Nefropatia cu leziuni glomerulare minime – alte GNC primitive

-edificatoare – punctia biopsie renala

GNC primitive – GNC de cauze secundare

Lipsa altor manifestari clinice si anamneza negativa pentru cauze infectioase (virus B,C, HIV, infectie cu streptococ beta-hemolitic grup A), colagenoze (LES, vasculite), neoplazii, nefrotoxice

DIAGNOSTIC POZITIV FINAL

Boala cronica de rinichi stadiul G1 KDIGO (RFG=96,8 ml/min/1,73 mp)

Glomerulonefrita cronica primitiva cu leziuni minime forma cu sindrom nefrotic activa

Hipertensiune arteriala secundara

Hipoalbuminemie secundara severa. Hipoproteinemie secundara severa

Dislipidemie mixta

Hipotiroidie subclinica pe fond de tiroidita cronica autoimuna

Insuficienta venoasa cronica de membre inferioare CEAP CII

Litiaza biliara multipla simptomatica

Steatoza hepatica

Anexita cronica stanga

Climax precoce

EVOLUTIE

- Grevata de complicatii
- Stadiile BCR

COMPLICATII

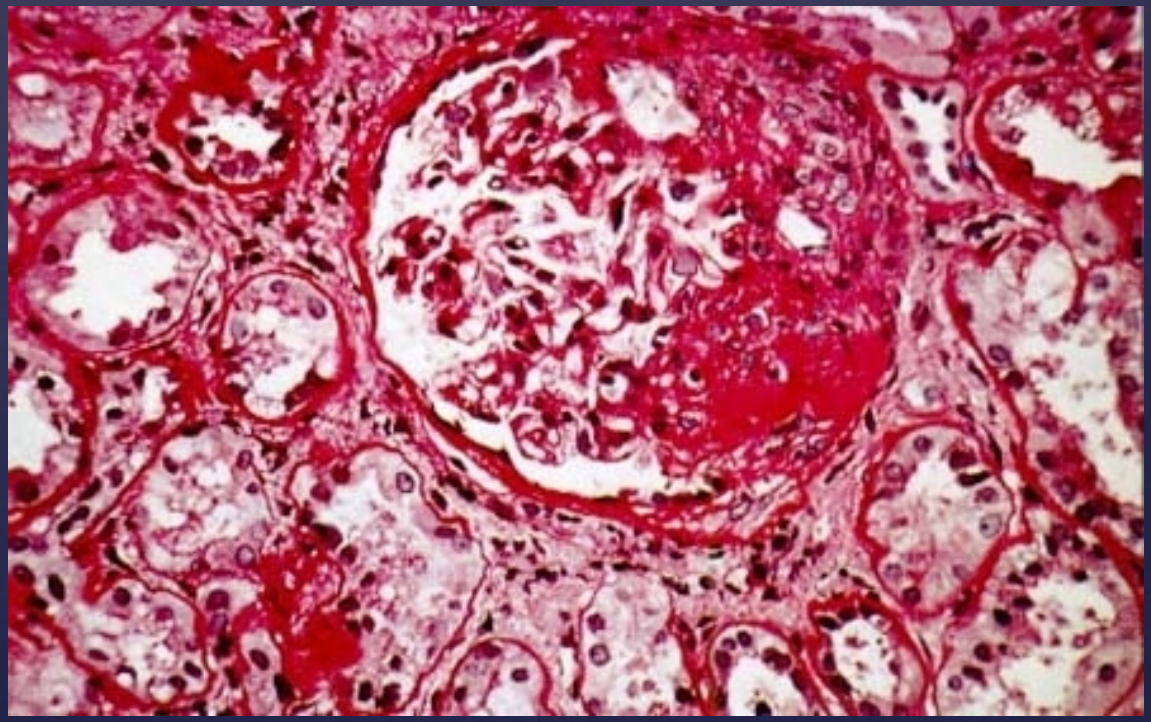
- HTA secundara
- Dislipidemie mixta
- Hipoalbuminemie, hipoproteinemie
- Hipotiroidie

- Infectioase
- Tromboze
- Embolii
- Sindrom Fanconi secundar disfunctiei tubulare proximale datorita proteinuriei nefrotice
- Ale tratamentului: osteoporoza, diabet zaharat, miopatie cortizonica

PROGNOSTIC

- Pe termen scurt: este dictat de elementele prognostice
  - - pozitive: functie renala buna, incadrabila ca stadiu incipient al BCR
  - - negative: HTA, hipoproteinemie, hipoalbuminemie, dislipidemie -> factori de risc CV

  - Pe termen lung:
  - Devine rezervat prin modalitatea de raspuns la tratamentul patogenic,
  - Pe PBR extensia de matrice, leziuni de depozite mezangiale -> leziuni de GSFS



TRATAMENT

- KDIGO 2012 Clinical Practice Guideline for Glomerulonephritis
- ►Treatment of initial episode of adult MCD
- ●corticosteroids be given for initial treatment of
- nephrotic syndrome.
- - prednisone or prednisolone be given at a daily single dose of 1 mg/kg (maximum 80 mg) or alternate-day single dose of 2 mg/kg (maximum 120 mg).
- - the initial high dose of corticosteroids, if tolerated, be
- maintained for a minimum period of 4 weeks if complete remission is achieved, and for a maximum period of
- 16 weeks if complete remission is not achieved. In patients who remit, we suggest that corticosteroids be tapered slowly over a total period of 6 months

TRATAMENT

- for patients with relative contraindications or intolerance to high-dose corticosteroids (e.g., uncontrolled diabetes, psychiatric conditions, severe osteoporosis), we suggest oral cyclophosphamide or CNIs as discussed in frequently relapsing MCD.
- the same initial dose of corticosteroids for infrequent relapses as in Recommendation 5.1.2 until remission is achieved, followed by at least 2 months of tapering steroids.

[KDIGO Clinical Practice Guideline for Glomerulonephritis. *Kidney Int* 2012; Suppl 2: 139–274]

TRATAMENT

- Frequent Relapses/Steroid Dependent MCD
- ●oral cyclophosphamide 2-2.5 mg/kg/d for 8 weeks.
- ●CNI (cyclosporine 3-5 mg/kg/d or tacrolimus 0.05-0.1 mg/kg/d
- in divided doses) for FR/SD MCD patients who have relapsed despite cyclophosphamide, and for people who wish to preserve fertility.
- ●MMF 750-1000 mg twice daily for patients who are intolerant
- of corticosteroids, cyclophosphamide, and CNIs.
- ►Corticosteroid-resistant MCD
- ●re-evaluate patients who are corticosteroid-resistant for other causes of nephrotic syndrome.

[KDIGO Clinical Practice Guideline for Glomerulonephritis. *Kidney Int* 2012; Suppl 2: 139–274

TRATAMENT

- Din 30.11.2014 - Prednison 5 mg 11 cp/zi ,1 luna -> reducere progresiva cu 5 mg/2 saptamani pana in 13.05.2014 – remisiune completa
- Simvastatin 40 mg 0-0-1
- Cal-D-Vita 1-0-0
- Pantoprazol 40 mg 1-0-0
- Lasix 500 mg 1/4-1/4-0
- Ramipril 2,5 mg 1/2-0-0
- ! Medicatie autosistata din mai 2015

TRATAMENT

- 02.2017: edeme gambiere, astenie, creatinina serica = 0,7 mg/dl, proteinurie= 1,02 g/24 ore

=> Aprovel 150 mg 0-0-1

- 07.2017: edeme generalizate, dispnee cu ortopnee, TA=180/100 mmHg creatinina serica = 0,7 mg/dl , proteinurie = 2,5 g/24 ore

=> X Prednison ?

X Ciclofosfamida ?

! Ciclosporina 3 mg/kgc/zi

Auto-sistare

- 11.2017: edeme declive, dispnee cu ortopnee

creatinina serica=0,9 mg/dl, proteinurie= 4,5 g/24 ore

Ciclosporina 3 mg/kgc/zi

Auto-sistare

- 07.2018: edeme declive, dispnee cu ortopnee, greturi, varsaturi

creatinina serica=0,76 mg/dl, albumine serice=1,6 g/dl, proteinurie=3 g/24 ore

Ciclosporina 3 mg/kgc/zi

TRATAMENT

- Dieta: Aport hidric oral: diureza+700 ml, Hiposodata – max 2g sare/zi, Hipolipidica 1 g/kgc/zi, Hipoproteica 0,8 g/kc/zi
- Scadere G: IMC=25 kcal/kg/zi
- Efort fizic adaptat
- Monitorizare: TA (tinta terap: 130/80 mmHg), FC, T, diureza,
  la 3 luni: HLG, uree, creatinina, sediment Addis, proteinurie/24 ore, urocultura
- Medicamentos:
- Neoral 50 mg 3-0-2
- Albunorm 100 ml/zi
- Furosemid fiole 2fi/12 ore, ulterior oral
- Irbesartan 150 mg 1-0-1
- Leridip 10 mg 1-0-1
- Sortis 20 mg 0-0-1
- Euthyrox 25 mcg 1+1/2-0-0
- Stabilirea momentului operator, avand in vedere patologia de fond (hipoalbuminemie – vindecare plagii)

PARTICULARITATEA CAZULUI

Depozitele mezangiale de IgM, C1q si C3

– elemente predictive pentru corticorezistenta/corticodependenta si suprapunerea leziunilor de GSFS