Bệnh Thận:			1185		
Thận hư	N04		550		46.41
Viêm cầu thận cấp	N00		183		15.44
Nhiễm trùng tiểu	N39.0		44		3.71
Henoch-Schonlein	D69.0		72		6
Lupus	L93		244		20.6
Suy thận cấp	N17		14		1.2
Suy thận man	N18		38		3.2
Bệnh Thận khác	_		40		3.44
Bệnh Nội tiết:			**121**		
Tăng sinh thượng thận	E25		**8**		6.6
Tiểu đường	E10		**54**		44.62
Cường giáp	E05		7		5.8

COMPLICATIONS OF NEPHROTIC SYNDROME: OEDEMA AND LOW PLASMA VOLUME

- **Gross oedema**
  - Fluid restriction to 500-1000 ml/day
  - Salt restriction
  - Frusemide 1-2 mg/kg/dose daily or twice daily or by continuous infusion
  - Albumin infusion (1-2 g/kg over 4-6 hrs) with frusemide
    - Risk of pulmonary oedema
  - Aldosterone antagonist (spironolactone) Use diuretics with caution if without albumin
  - Thiazide diuretics or metolazone

Acute renal failure and shock

- Caused by low plasma volume, diuretics, additional fluid losses in gastroenteritis, cyclosporin
- Resuscitate with 0.9% saline, albumin(0.5 mg/kg) over 6-8 hours (no diuretic)

EMERGING THERAPIES

1. TGFβ
2. TNF
3. JAK/STAT
4. B7.1
5. Retinoic acid
6. Anti-complement
7. Notch1
8. Anti-suPAR

- **Sang thương tối thiểu hay tăng sinh trung mô lan tỏa có thể cho:**

  Cyclophosphamide và Prednisone

FSGS

BỊNH CẦU THẬN MANG

VIEM CẦU THẬN TĂNG SINH MANG

MCNS, MESP-GN, FSGS :

Cylosporine: 5mg/kg/ngày

Hay Tacrolimus 0,15 mg /kg chia 2

Prednisone:

1mg/kg/cách ngày x 5 tháng

Tacrolimus vs cyclosporin- observational study- Wang 2012. No difference in relapse rate

MMF vs CSA- RCT. Higher relapse rate with MMF

MMF vs CSA crossover design (Gellerman)- relapse free rate on CSA 84%; on MMF 64%

Agent	N of RCT's	N=	Risk ratio of relapse	Time of outcome mths	Rel risk reduction
Cyclophosphamide	3	102	0.44 (0.26,0.73)	6-12	56%
Levamisole	5	269	0.43 (0.27,0.68)	4-12	57%

EFFECTIVE STEROID SPARING AGENTS FOR SSNS

Cyclophosphamide	2 mg/kg/day	8-12 weeks
Chlorambucil	0.1-0.2 mg/kg/day	8-12 weeks
Levamisole	2.5 mg/kg on alt days	12 months or more
Cyclosporin*	4-5 mg/kg/day in 2 doses	12 months or more
Tacrolimus*	0.1 mg/kg/day in 2 doses	12 months or more
Mycophenolate mofetil	1200 mg/m^2/day in 2 doses	12 months or more
Rituximab	375 mg/m^2 per dose	?once /once yearly as required

* Starting dose; monitor levels

ĐIỀU TRỊ TÁI PHÁT

Tái phát thường xuyên, hoặc lệ thuộc corticoid:

Prednisone 2mg/kg/ngày cho đến khi đạm niệu (-) 3 ngày liên tiếp

Sau đó : Prednisone 1,5 mg/kg/ cách ngày, trong 4 tuần

Tiếp theo giảm liều dần, rồi duy trì: 0,1-0,5mg/kg/cách ngày trong 3-12th

TPTX : 3-6th

Phụ thuộc : 9-12th

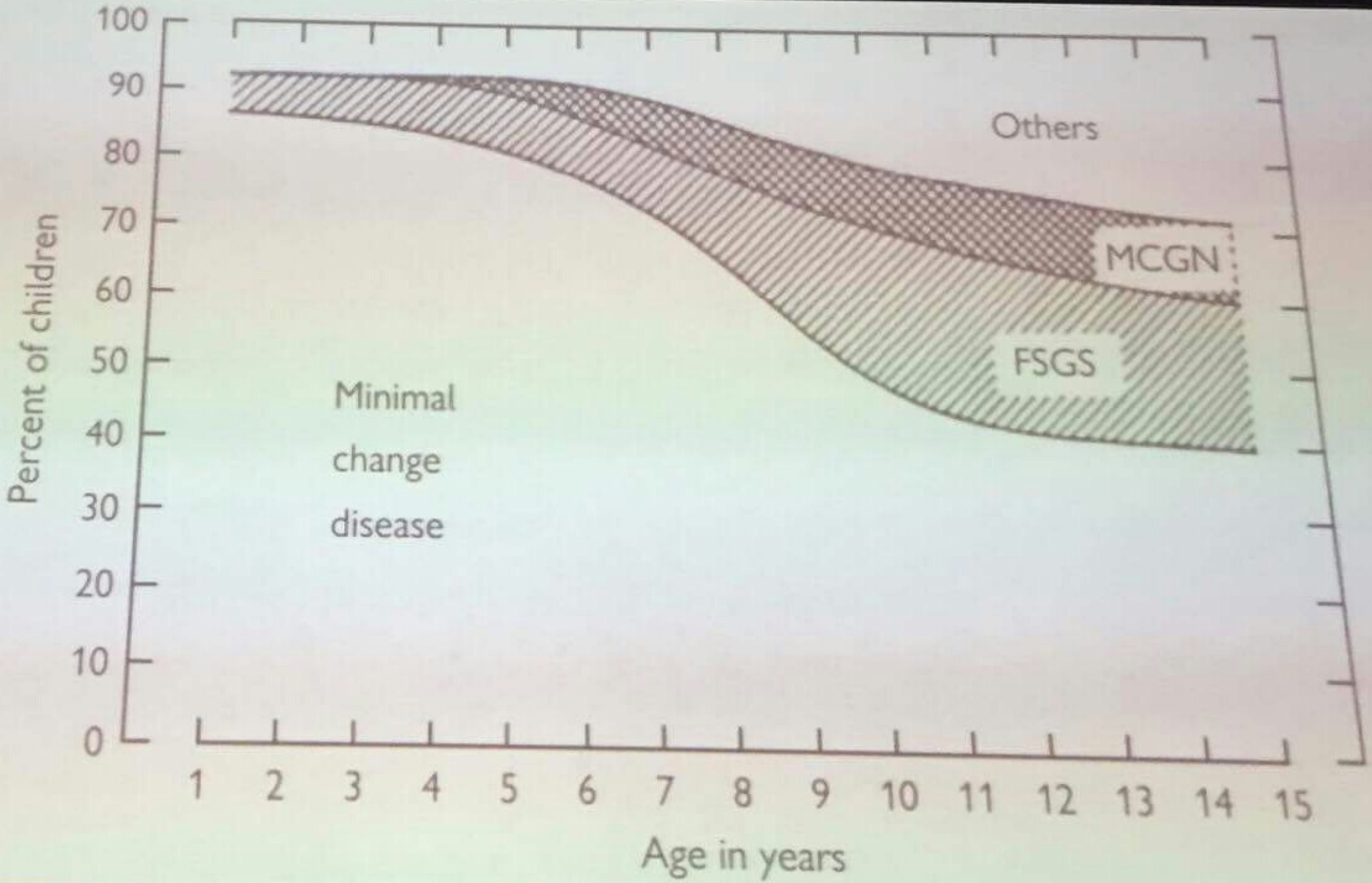
- **Nếu lúc giảm liều bị tái phát với liều prednisone > 0,5 mg/kg/ cách ngày → Cho thêm: Levamisole 2,5 mg/kg/cách ngày, trong 4 - 12 tháng.**

2015 : 2 th – 3 th

4 W TẤN CÔNG : 2mg/kg/ ng

4 W CÁCH NGÀY : 1,5 mg /kg / cách ngày

4 W GIẢM LIỀU


100
90
80
70
60
50
40
30
20
10
0
Percent of children
Others
MCGN
FSGS
Minimal
change
disease
1
2
3
4
5
6
7
8
9
10
11
12
13
14
15
Age in years

PENICILLIN PROPHYLAXIS

- No controlled trials of use in NS
- 1 trial in sickle cell children- reduced the incidence of invasive pneumococcal disease from 16% to 2%. (Gaston MH 1988)
- Analysis: (P. McIntyre 1998) If assume invasive pneumococcal disease incidence is 1% and relative risk reduction of 80% then need to treat 110 NS children to prevent 1 episode of pneumococcal disease
- Conclusion: Not routinely recommended- but there is a low side effect profile- and would use if high risk population or unvaccinated

COMPLICATIONS OF NEPHROTIC SYNDROME: INFECTIONS

- **Bacterial infections**
  - Peritonitis, septicaemia, cellulitis, pneumonia, UTI
  - *S. pneumoniae*, *Haemophilis influenzae b*, *E.coli*
  - Risk factors
    - low serum IgG, low factors B & D, abnormal T cell function
    - immunosuppressive therapies
  - Reduce risk by
    - Immunisation against pneumococcus (Prevenar, Pneumovax)
    - Immunisation against *Haemophilis influenzae b*
    - Antibiotic prophylaxis with penicillin
- **Viral infections**
  - Varicella in immunosuppressed children; Vaccine, VZ IgG, aciclovir/valaciclovir

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RITUXIMAB SDNS 6 17

Franz Schaefer (Heidelberg, Germany) reported that a single course of rituximab is significantly more effective than 12 months tacrolimus in maintaining disease remission in children with steroid-dependent nephrotic syndrome (SDNS), according to a study from India. After 12 months, 90 % of children receiving rituximab were relapse free compared with 63 % of the tacrolimus group ($p < 0.001$). Children treated with rituximab also needed significantly lower steroid doses, and had better growth, and better kidney function. Professor Schaeffer concluded that, given its excellent tolerability, rituximab may be considered as first-line steroid-sparing therapy in children with SDNS. The open-label, single-center, parallel-arm trial randomized 120 children with SDNS either to 12 months tacrolimus plus tapering doses of alternate day prednisolone or to a single course of rituximab. Tacrolimus was dosed at 0.2 mg/kg/day targeting 5 – 7 ng/ml trough level, while rituximab was given as two to four weekly infusions at 375 mg/m2 depending on circulating B-cell count.

Long-term efficacy and safety of common steroid-sparing agents in idiopathic nephrotic children

Biswanath Basu[1] · Binu George Babu[1,2] · T. K. S. Mahapatra[3]

Received: 10 March 2016 / Accepted: 31 March 2016

Abstract

Background Calcineurin inhibitors (CNI), mycophenolate mofetil (MMF), and levamisole are common treatment choices for patients with frequently relapsing (FRNS) and steroid-dependent nephrotic syndrome (SDNS).

Methods In this retrospective cohort study, we analyzed the relative efficacy and safety of tacrolimus, MMF, and levamisole over a period of 30 months, in treating 340 children with idiopathic FRNS/SDNS. The children received either MMF 1200 mg/m^2 daily, or levamisole 2.5 mg/kg on alternate days, or tacrolimus 0.1–0.2 mg/kg daily along with tapering doses of alternate-day prednisolone.

Results Tacrolimus was associated with a higher rate of 30-month relapse-free survival when compared to MMF (61.7 vs. 38.5 %, $p < 0.001$), or levamisole (61.7 vs. 24 %, $p < 0.001$). However, relapse rate increased almost three-fold once tacrolimus was stopped, resulting in a higher relapse rate per patient-year when compared to the MMF group (2.0 vs. 1.5, $p = 0.013$). The cumulative pred-relapse risk was higher in patients with steroid dependency at baseline (HR 2.14, 95 %CI 1.79–2.96, $p < 0.0001$). In comparison with few minor adverse events in other two cohorts, several serious adverse events were documented in the tacrolimus group.

Conclusions Although there are serious safety concerns regarding tacrolimus, it is more effective than MMF or levamisole in maintaining relapse-free survival. However, unlike MMF, the relative efficacy of tacrolimus in preventing further relapses is seen only when the patient is on the drug. Taking together the long-term efficacy and safety data observed, MMF appears as a safe and effective alternative to tacrolimus in managing pediatric FRNS/SDNS.

Keywords Nephrotic syndrome · Mycophenolate mofetil · Levamisole · Tacrolimus

Introduction

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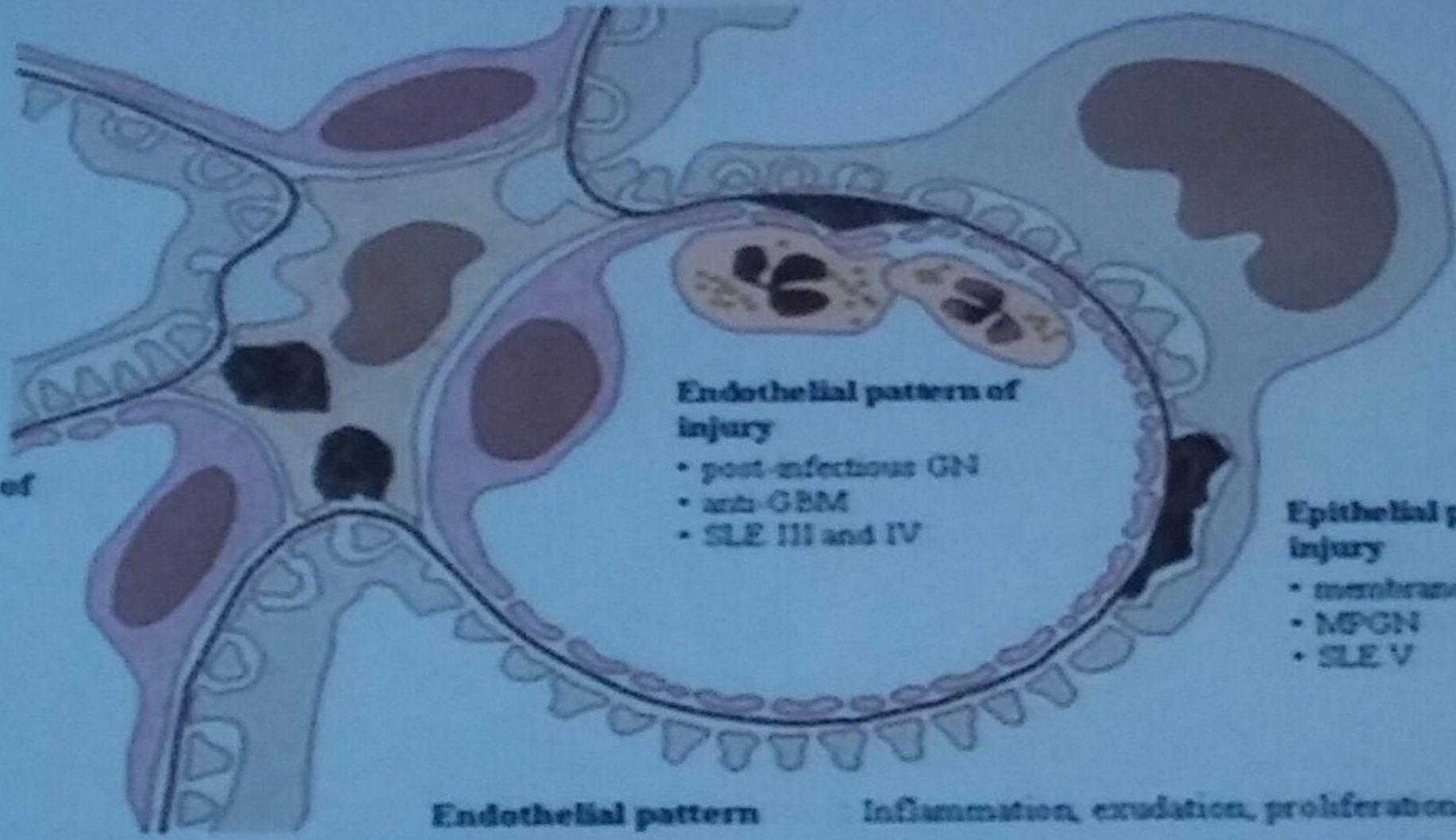
Glomerular injury determined by immune complex localization



mesangial pattern of injury
- IgA nephropathy
- MPGN
- SLE II

Endothelial pattern of injury
- post-infectious GN
- anti-GBM
- SLE III and IV

Epithelial pattern of injury
- membranous GP
- MPGN
- SLE V

Endothelial pattern	Inflammation, exudation, proliferation, GFR ↓
Epithelial pattern	Non-inflammatory lesion, proteinuria
Mesangial pattern	Mesangial cell proliferation, hematuria

- **Sang thương tối thiểu hay tăng sinh trung mô lan tỏa có thể cho:**
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FSGS
BỆNH CẦU THẬN MANG
VIEM CẦU THẬN TĂNG SINH MANG