

# CRIMEAN FEDERAL UNIVERSITY

## Works on E.N pavlovsky-natural focal diseases

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# Who is E.N PAVLOVSKY?

- ▶ Yevgeny Nikanorovich Pavlovsky (Russian: Евге́ний Никано́рович Па́вловский; 22 February (N.S. 5 March) 1884, today's Voronezh Oblast – 27 May 1965, Leningrad)
- ▶ He was a Soviet zoologist, entomologist, academician of the Academy of Sciences of the USSR (1939), the Academy of Medical Sciences of the USSR (1944), honorary member of the Tajik Academy of Sciences (1951), and a lieutenant-general of the Red Army Medical Service in World War II.

# History

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- Recent upper respiratory (1-2wks back), skin infection (3-6 wks): PSGN or GI infection: HUS, HSP nephritis
- H/o ingestion of drugs (ATT – Rifampicin, Ibuprofen, Chloroquine, Metronidazole, Iron), i.v. contrast agents (Toxic nephropathy, RVT)

# What is focal disease?

## **Focal Disease**

- Patients with focal disease have a less favorable prognosis. In a study of 530 patients (including 170 patients with focal disease); those with focal disease had a greater likelihood of therapeutic failure, relapse, or death.

# Background

- Disease “entity” defined by findings on the kidney biopsy
- Characterized by **scarring or hardening** of *glomeruli*
  - Affect alteration of normal glomerular structure and function
- It is a major cause of idiopathic steroid-resistant nephrotic syndrome in children and adults
- Unlike minimal change disease, FSGS often progress to end-state renal disease (ESRD)

# Theory

## Focal Infection Theory

- ❑ Good example of how research changes
- ❑ A very popular theory in the 1920s and 30s, thought that many diseases originated in the mouth
- ❑ Millions of healthy teeth were extracted, until the theory was discredited
- ❑ Since the 1990s, new research has been showing oral-systemic connections
- ❑ Where will this go?



# Cause

- **Primary-**

- Minimal change disease- commonest in children- CRI ~10-20%
- Focal segmental glomerulosclerosis- CRI ~50%
- Membranous nephropathy- commonest in adults- CRI ~1/3<sup>rd</sup>

- **Secondary-**

- DM
- SLE
- HIV infection-FSGS, viral hepatitis, malaria
- Amyloidosis
- Sarcoidosis
- Drugs- NSAIDs, gold, penicillamine
- Cancer- Hodgkin's disease, NHL, solid tumors-GIT, RCC, lung

# Distribution of disease

## CLASSIFICATION OF GLOMERULAR DISEASE BY DISTRIBUTION

*A) Classification of disease distribution when many glomeruli are considered.*

**FOCAL** - Disease affecting only some of glomeruli.

**DIFFUSE** - Disease affecting most or all glomeruli.

*B) Classification of disease distribution when single glomeruli are considered.*

**SEGMENTAL** - a lesion involving only a part of the glomerulus

**GLOBAL** - a lesion involving the entire glomerulus.



# Heck's disease

## Focal epithelial hyperplasia (Heck's disease)

- It's also a human papilloma virus related disease , however this is seen in subset of people (native Indians), so its diagnosis is easier, also it's multiple and it's not localized , they occur in early age, they regress on their own (don't need any treatment), later on in life they go away on their own, they are not very obvious, you don't really want to worry about the management of this hereditary disease
- They are associated with:  
Koilocytes, thick epithelium,  
and related to HPV



## Focal epithelial hyperplasia Heck's disease

- ❑ Clinical features:
  - ❑ - Multiple nodular soft tissue masses
  - ❑ - Buccal, labial mucosa and tongue
  - ❑ - Whitish to pinky in color



**Focal epithelial hyperplasia of the buccal mucosa.**



## Focal fatty liver changes

- Focal fatty liver infiltration.



- Focal fatty sparing

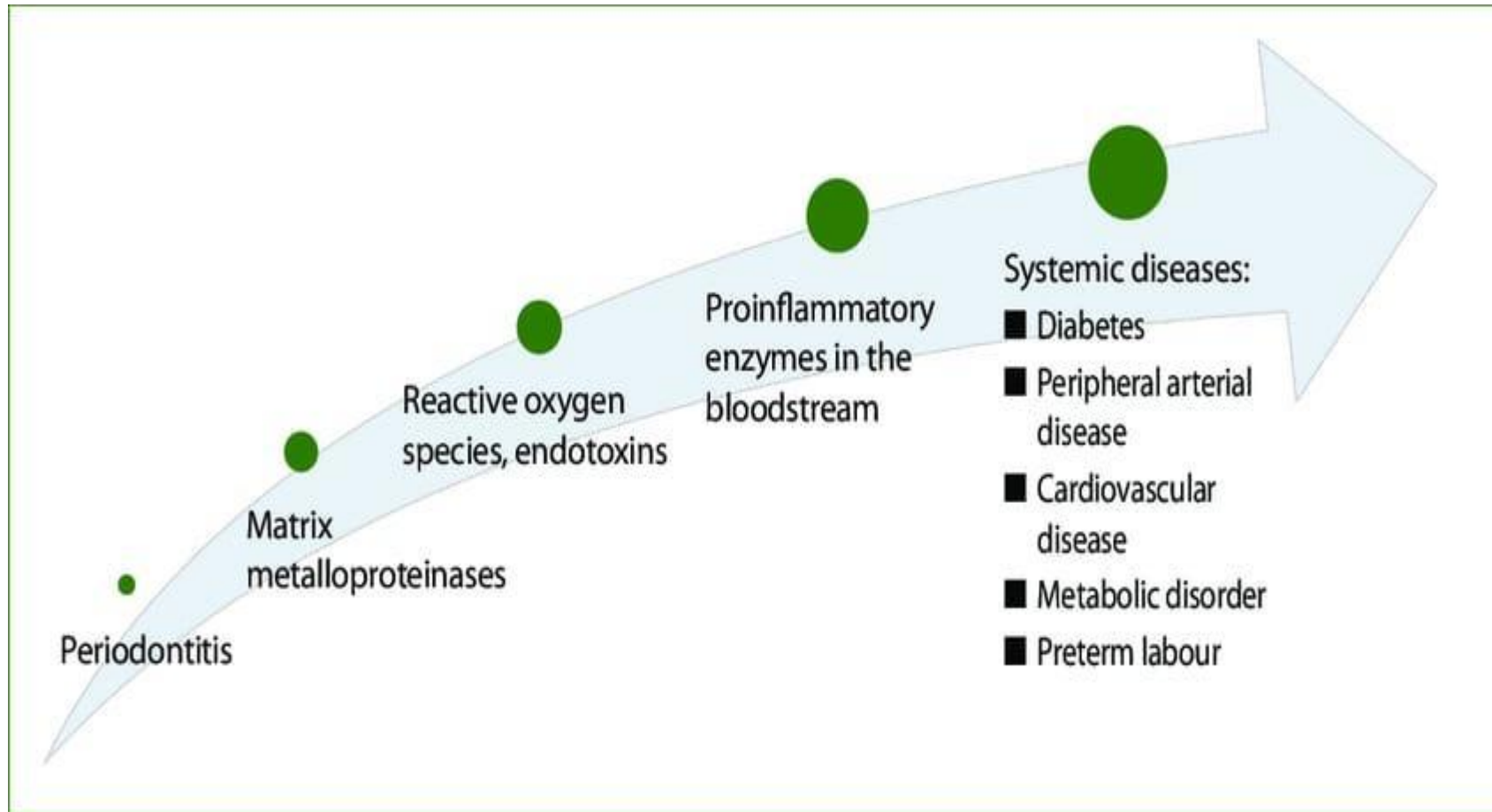


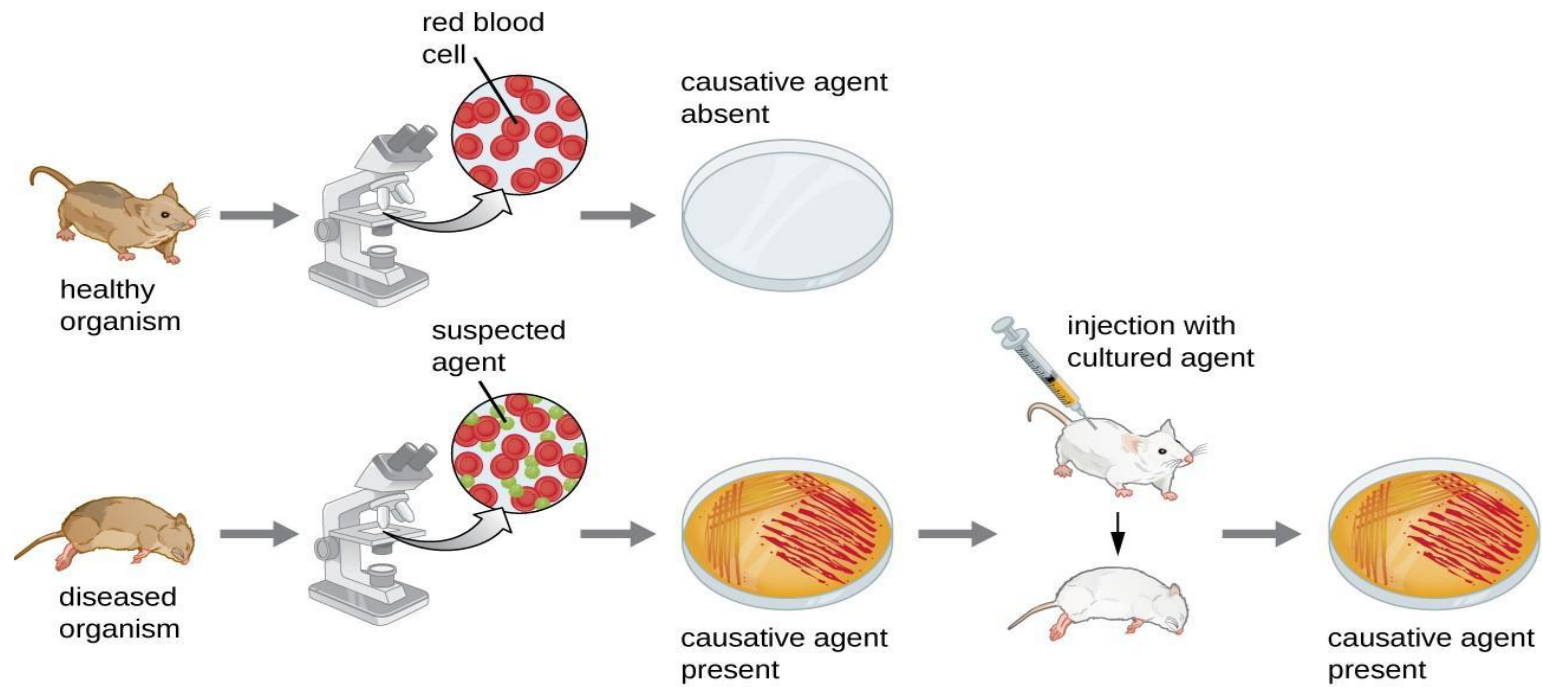
# Epidemiology

- Frequency:
  - Adults > children
- Prevalence in adults: **≥45** years old
- In US, >5400 patients are diagnosed with FSGS each year and ~**20K** patients are currently living with ESRD due to FSGS
- In children
  - The **2<sup>nd</sup> leading** cause of renal failure
  - Accounts for 15%-20% of cases with nephrotic syndrome
  - Most common cause of steroid-resistant nephrotic syndrome



# Disease steps





1 The suspected causative agent must be absent from all healthy organisms but present in all diseased organisms.

2 The causative agent must be isolated from the diseased organism and grown in pure culture.

3 The cultured agent must cause the same disease when inoculated into a healthy, susceptible organism.

4 The same causative agent must then be reisolated from the inoculated, diseased organism.

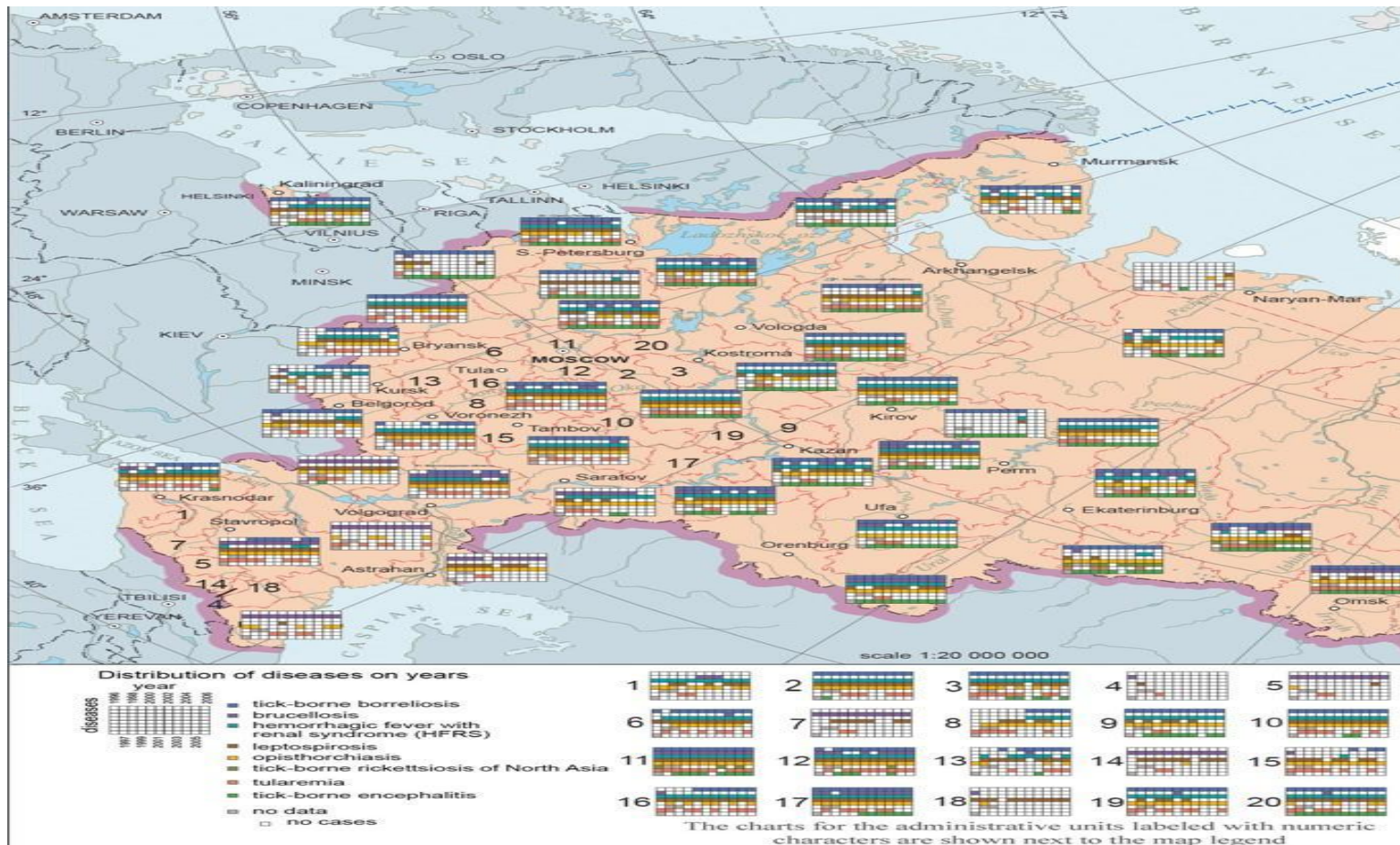
# Pathogenicity

- Key factor in the pathogenesis
  - **Podocyte damage and loss**
- Injury to podocyte occurs by 4 major mechanisms:
  - Alteration of the components of the slit diaphragm or interference with its structure
  - Dysregulation of the actin cytoskeleton
  - Alteration of the glomerular basement membrane or its interactions with the podocyte
  - Alteration of the negative surface charge of the podocyte

# Diagnosis result

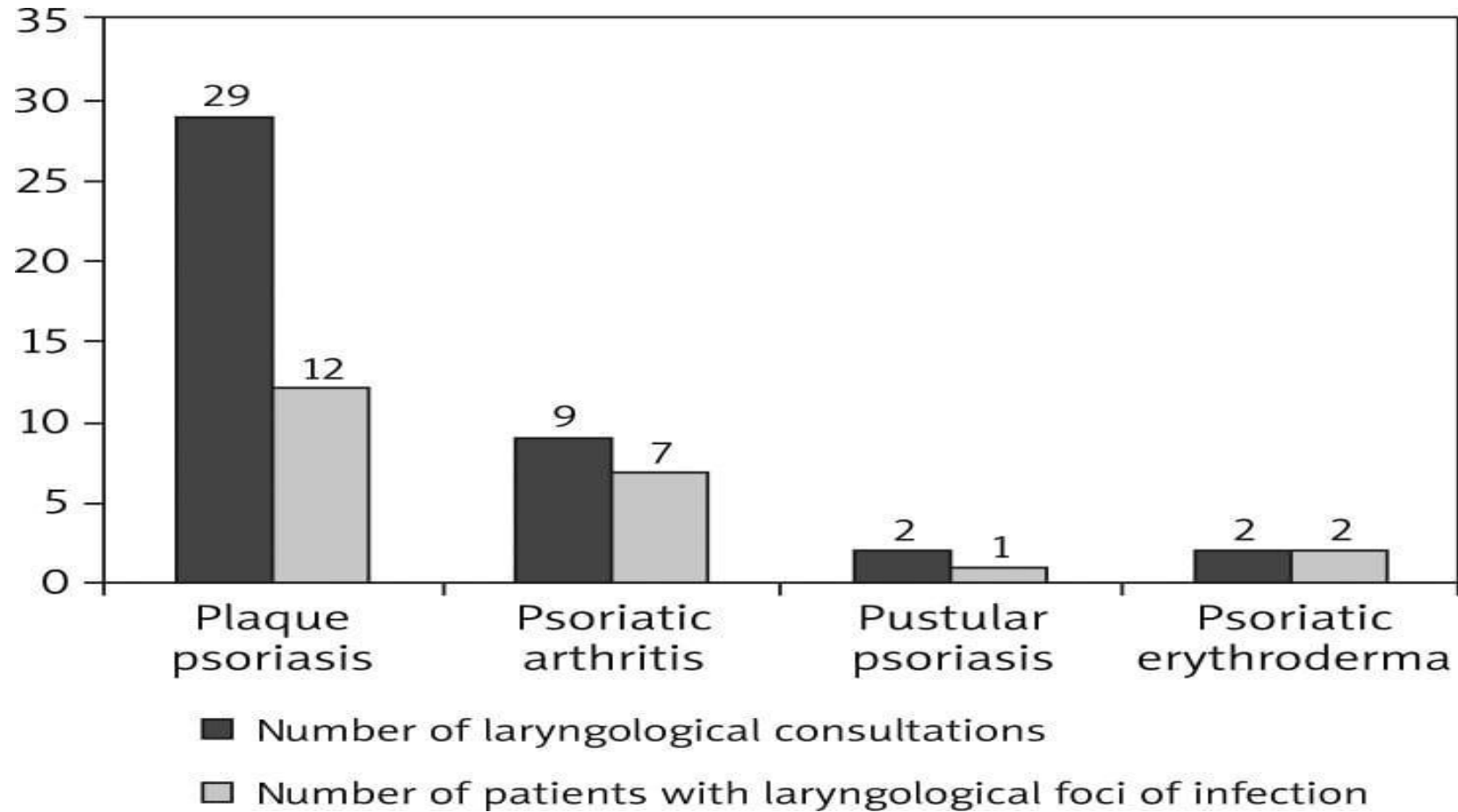
	<i>Focal malignant disease</i>	<i>Focal benign disease</i>	<i>Diffuse disease</i>
<i>Biopsy diagnosis</i>	<i>not confirmed at follow up</i>		
Lung tissue with no pathological changes	26	9	15
Necrotic tissue	5	4	0
Lung tissue with non-specific inflammation	0	3	5
Non-specific pulmonary fibrosis	1	0	1
No tissue	12	1	1
Biopsy lost	1	1	1
Total	45	18	23

# Maps of focal disease





# Graphs of disease



Thank  
you