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 Павло́вский; 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

- 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?

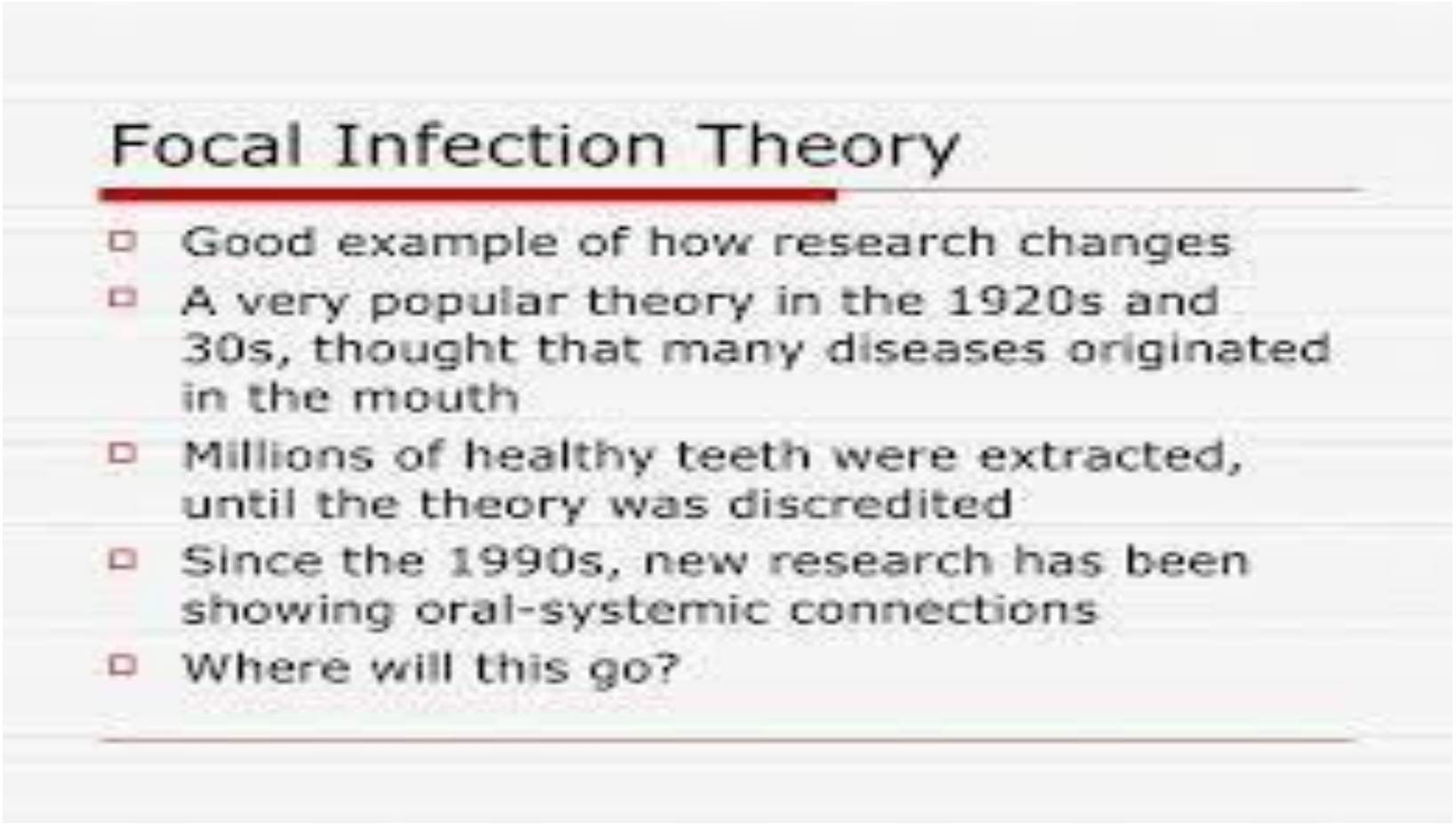
Focal Disease

- 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 alternation 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 ~1/3rd
- 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

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.

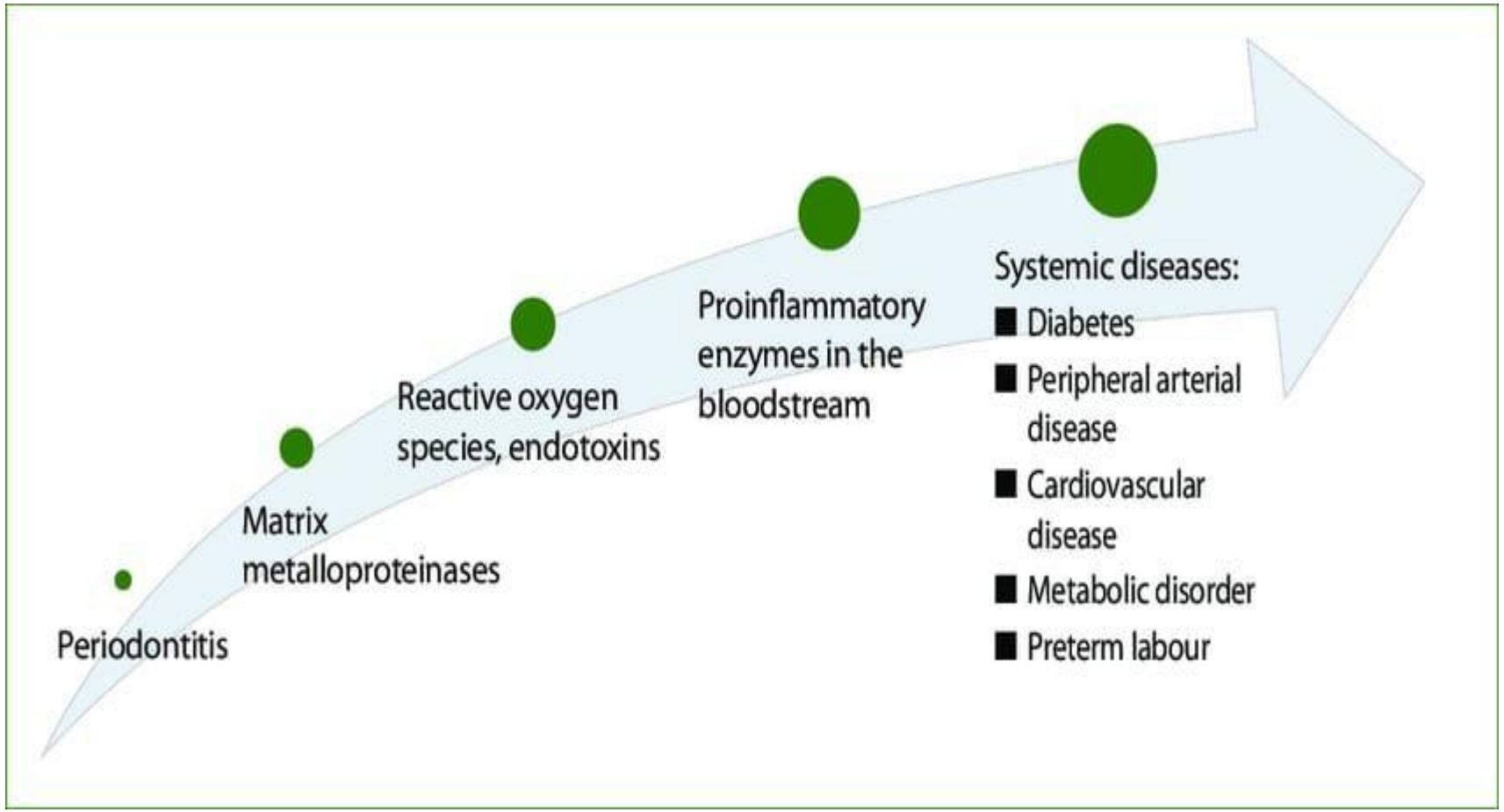
- Focal fatty sparing

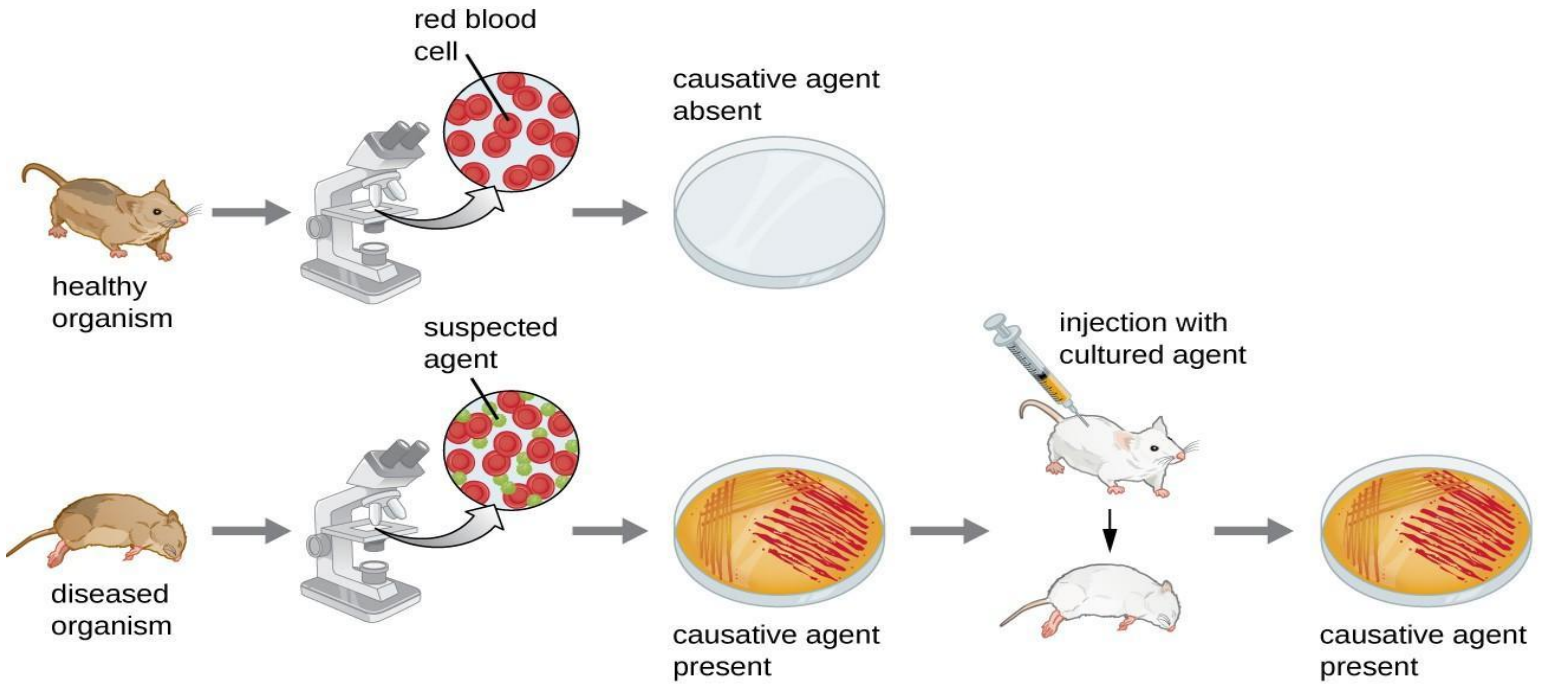
Epidemiology

- 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 2nd leading cause of renal failure
  - Accounts for 15%-20% of cases with nephrotic syndrome
  - Most common cause of steroid-resistant nephrotic syndrome

FSGS Facts. Accessed February 12, 2014. Available at: http://[illegible]

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

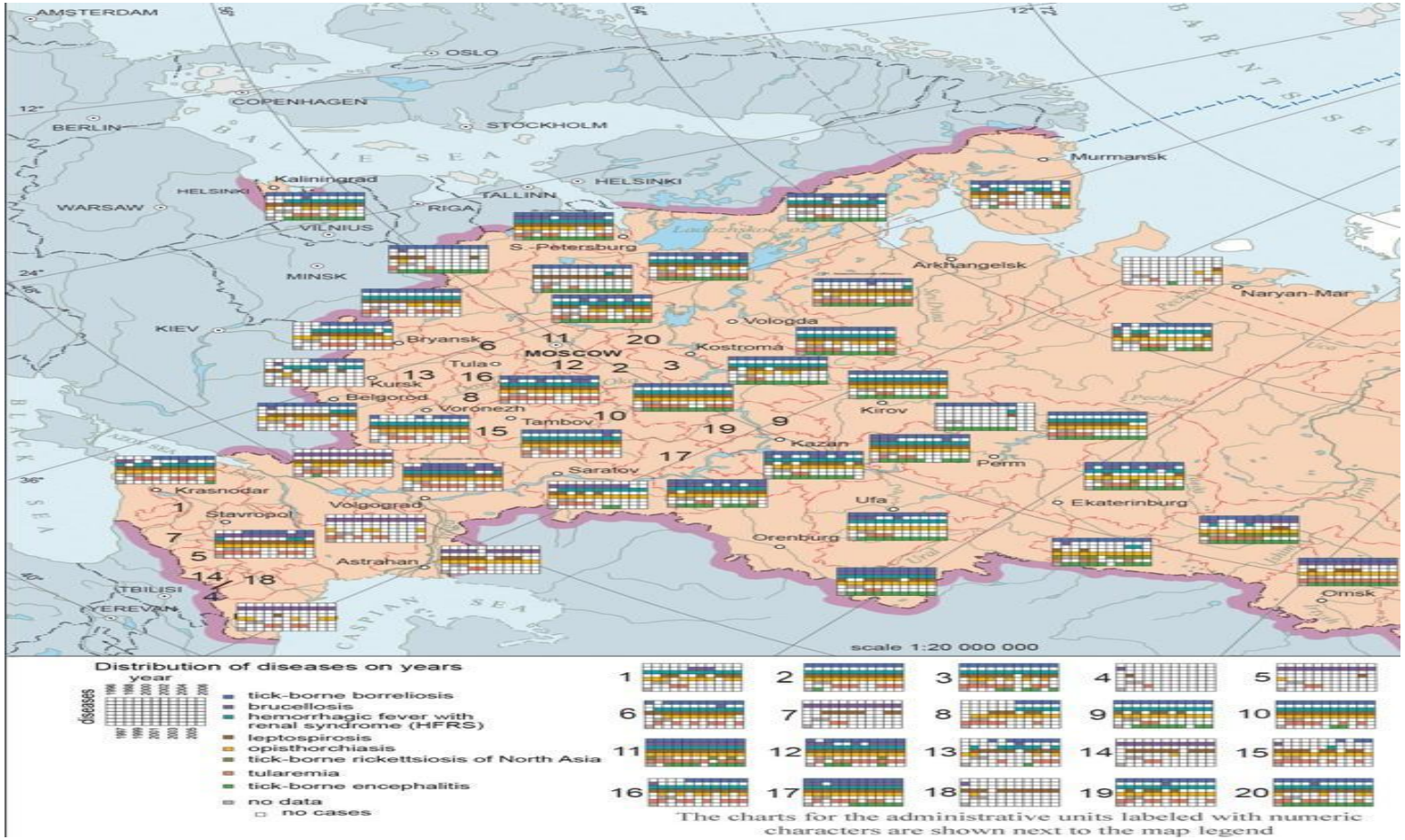
- Key factor in the pathogenesis
  - "**Podocyte damage and loss**"

- Injury to podocyte occurs by 4 major mechanisms:
  1. Alteration of the components of the slit diaphragm or interference with its structure
  2. Dysregulation of the actin cytoskeleton
  3. Alteration of the glomerular basement membrane or its interactions with the podocyte
  4. Alteration of the negative surface charge of the podocyte

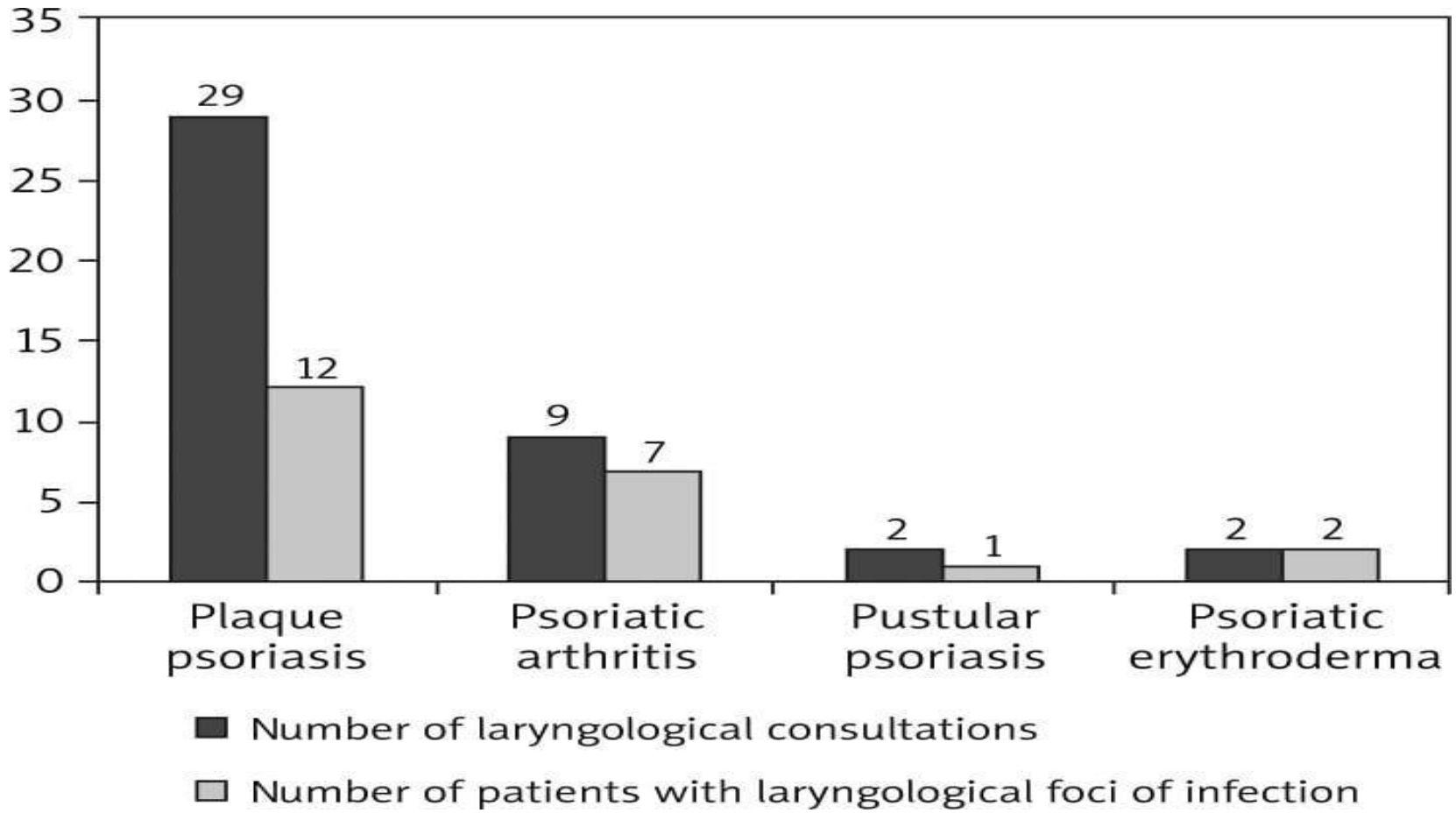
Diagnosis result

Biopsy diagnosis	*Focal malignant disease*	*Focal benign disease*	*Diffuse disease*
	not confirmed at follow up		
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
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