

STIKES NOTOKUSUMO  
YOGYAKARTA

# PATOFISIOLOGI SISTEM IMUN

*Presented by: Maria Putri Sari Utami, M.Kep.*

KEPERAWATAN DEWASA 2  
PRODI NERS



# JENIS PATOFISIOLOGI SISTEM IMUN

01

IMUNODEFISIENSI

02

AUTOIMUN

03

HIPERSENSITIFAS



# IMUNODEFISIENSI



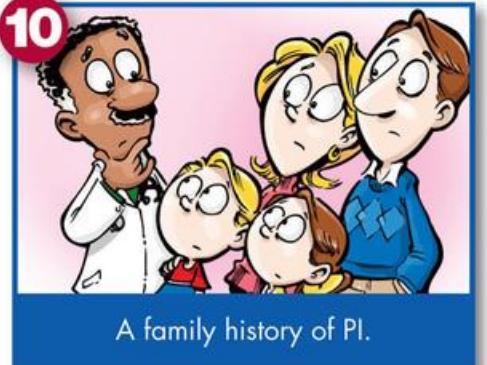
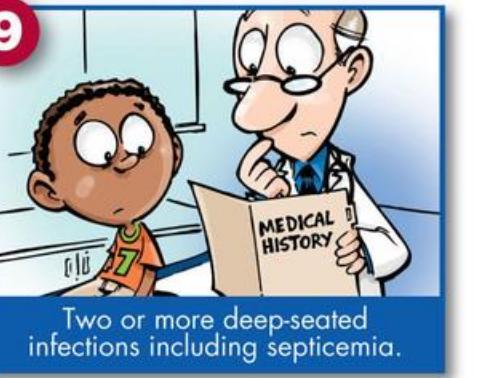
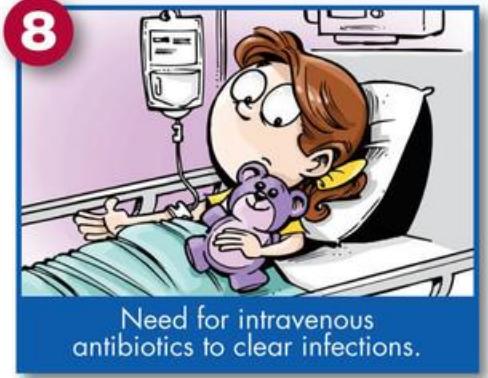
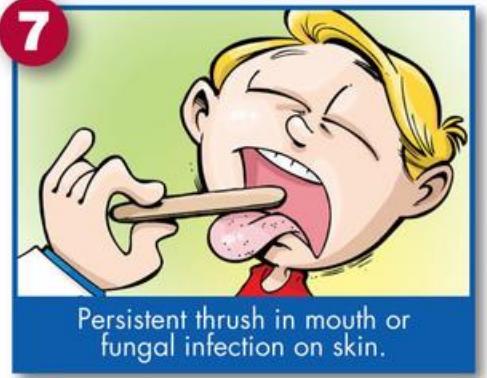
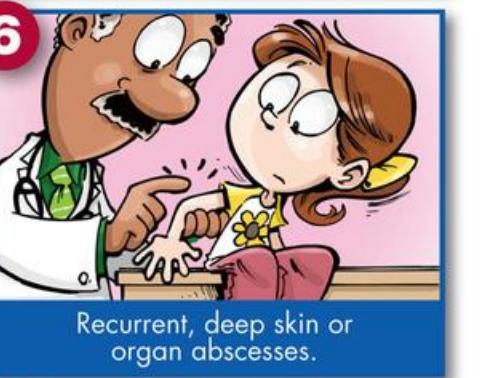
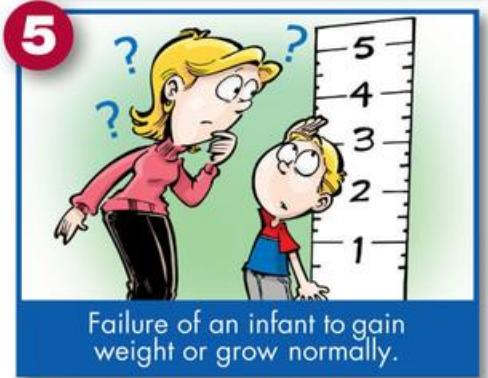
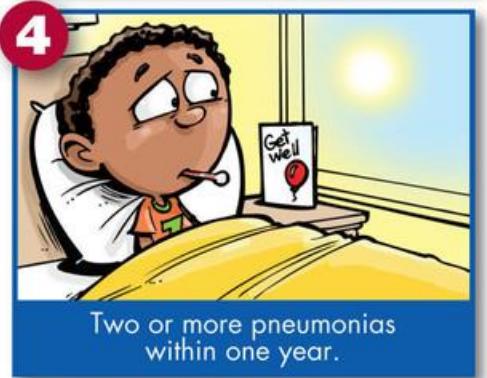
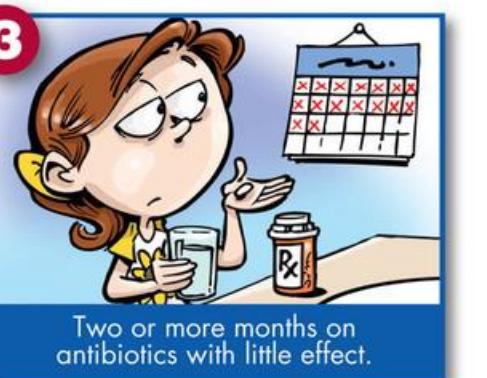
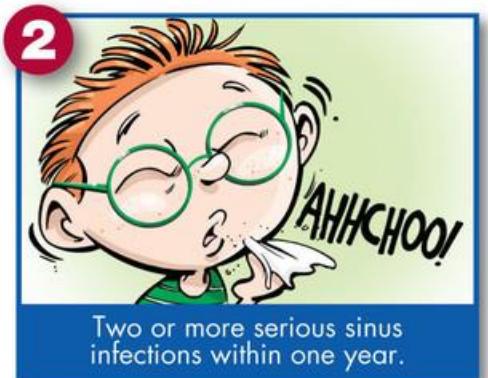
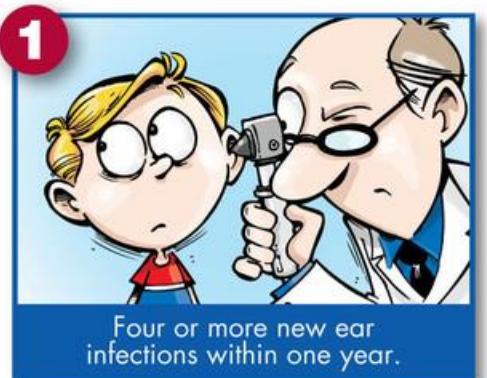
Imunodefisiensi merupakan penyakit yang disebabkan oleh hilang atau terjadinya gangguan fungsi dari sebagian sistem imun termasuk sel imun, molekul imun, atau proses kerja sistem imun.



Patofisiologi imunodefisiensi dari sangat bervariasi dan kompleks, menimbulkan gejala klinis yang banyak berbeda untuk setiap kejadian, megakibatkan penyakit serius

# 10 Warning Signs of Primary Immunodeficiency

Primary Immunodeficiency (PI) causes children and adults to have infections that come back frequently or are unusually hard to cure. 1:500 persons are affected by one of the known Primary Immunodeficiencies. If you or someone you know is affected by two or more of the following Warning Signs, speak to a physician about the possible presence of an underlying Primary Immunodeficiency.



Presented as a public service by:



Jeffrey Modell  
Foundation



Curing PI  
Worldwide



CDC

Funding was made possible in part by a grant from the U.S. Centers for Disease Control and Prevention (CDC).



NATIONAL  
CANCER  
INSTITUTE



PPTA



NIADDK



NICHD



Baxter  
BioScience



Bioteest  
From Nature for Life



CSL Behring  
Bioterapies for Life™



GRIFOLS



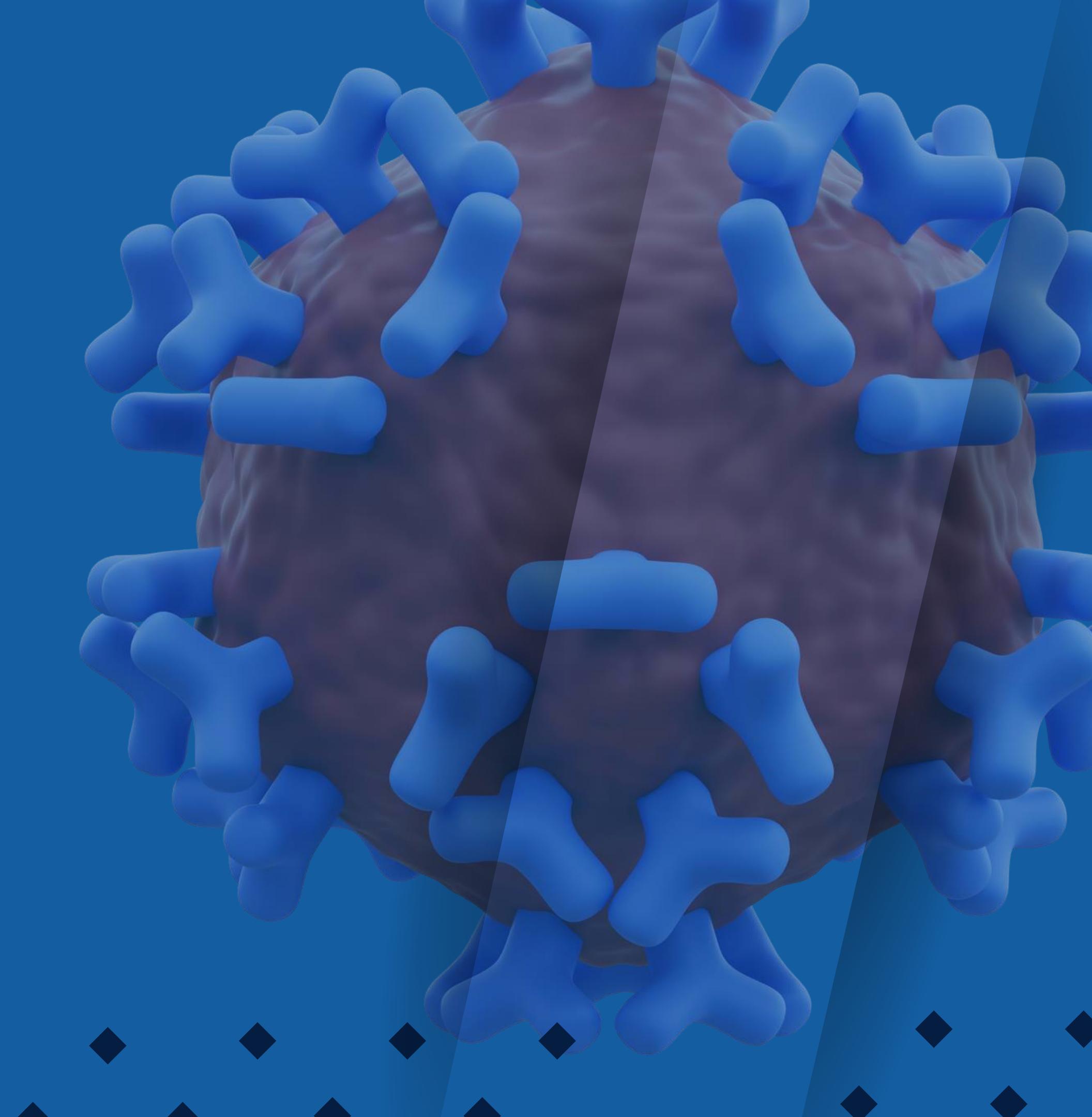
KEDRION  
BIOPHARMA



octapharma

These warning signs were developed by the Jeffrey Modell Foundation Medical Advisory Board.  
Consultation with Primary Immunodeficiency experts is strongly suggested. © 2013 Jeffrey Modell Foundation

For information or referrals, contact the Jeffrey Modell Foundation: [info4pi.org](http://info4pi.org) | 866-INFO-4-PI



# IMMUNODEFISIENSI



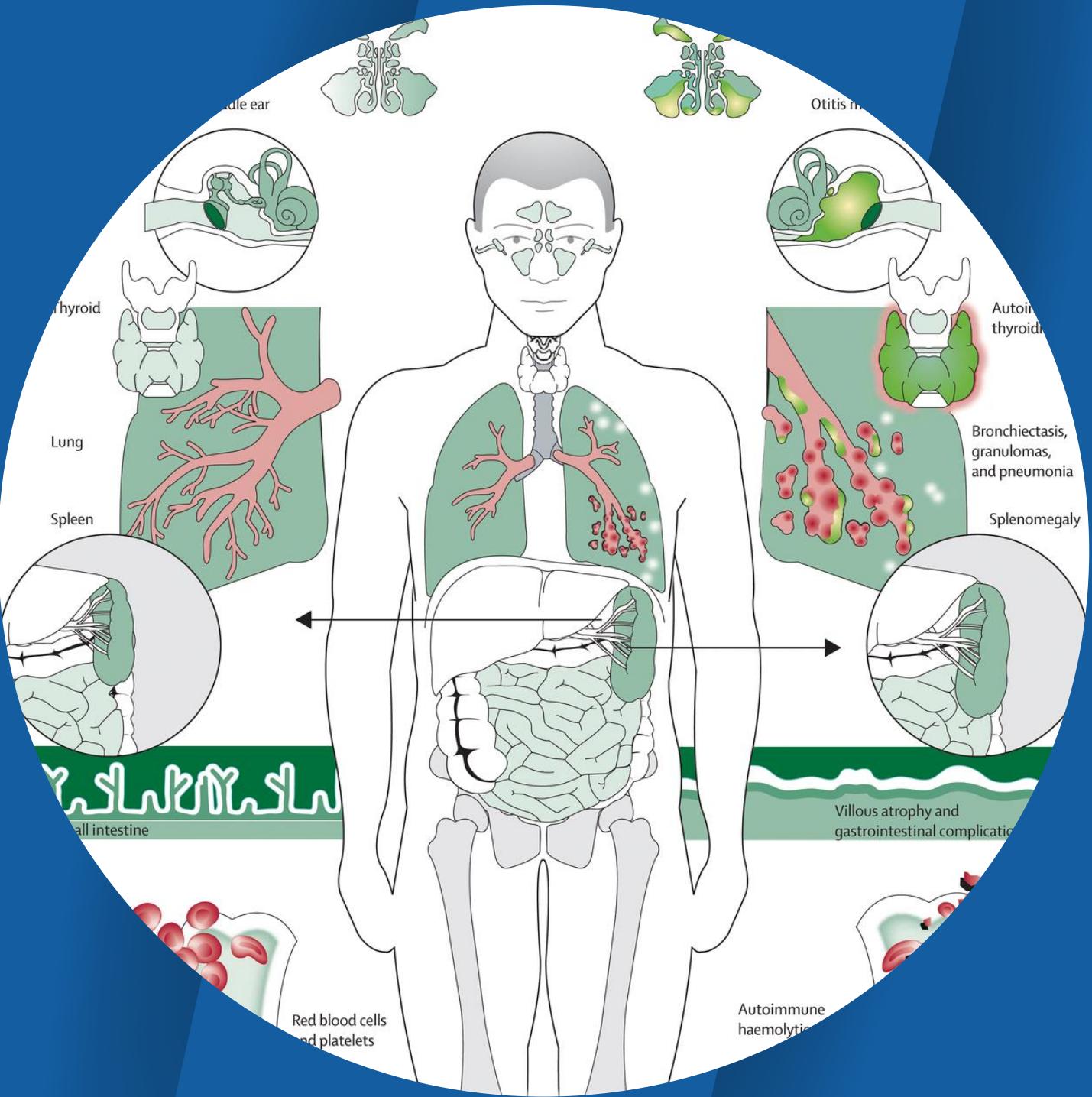
## PRIMARY

- < 10 %
- Intrinsic
- Missing enzyme (ADA)
- Missing cell type
- Nonfunctioning component
- Congenital
- Manifest since early age



## SECONDARY

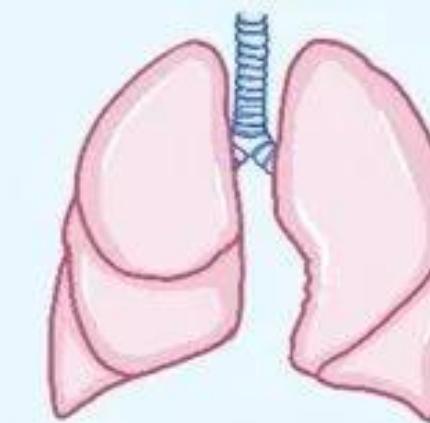
- 90 %
- Underlying disease
- Lymphoid malignancy
- HIV Infection
- Malnutrition
- Immunosuppressant drug
- Acquire
- Manifest in any age



## 2+ in 1 YEAR

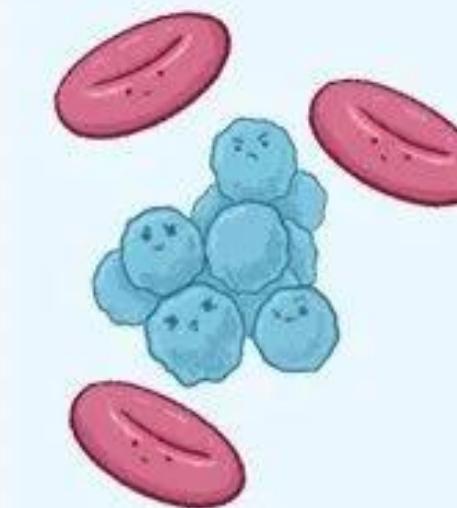


SERIOUS SINUSITIS

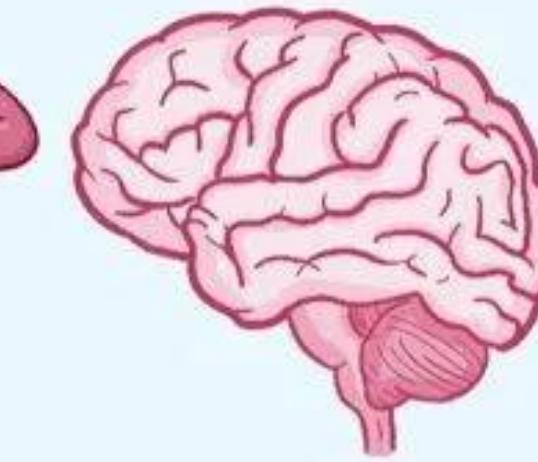


SERIOUS PNEUMONIA

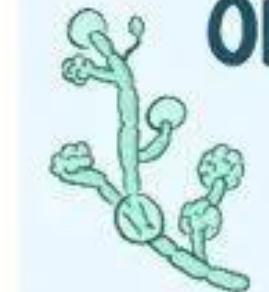
## 2+ in LIFETIME



SEPSIS



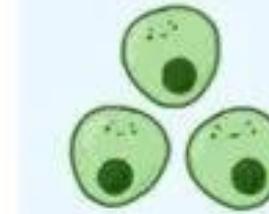
MENINGITIS



*Candida albicans*



nontuberculous  
Mycobacteria



*Pneumocystis jiroveci*

- ~ PERSISTENT FEVER
- ~ CONFINEMENT to BED for 1 WEEK+
- ~ DIFFICULT to TREAT
  - ↳ 2+ MONTHS of ANTIBIOTICS
  - ↳ IV ANTIBIOTICS
  - ↳ HOSPITALIZATION
- ~ UNUSUAL COMPLICATIONS

- ~ ORGAN ABSCESSES
- ~ NON HEALING WOUNDS
- ~ CHRONIC DIARRHEA
- ~ FAILURE to THRIVE
- ~ PERSISTENT LAB ABNORMALITIES
  - ↳ LEUKOCYTOSIS
  - ↳ ↑ ESR & CRP
- ~ PERSISTENT IMAGING ABNORMALITIES
  - ↳ BRONCHIECTASIS



# CAUSES OF SECONDARY IMMUNODEFICIENCY

Uremia, diabetes, malnutrition

**Metabolic**

**Iatrogenic**

Cytostatics, immunosuppressants

**Malignant tumors**

**Viral Infections**

HIV, CMV, measles, infectious mononucleosis

**Splenectomy**

**Stress**

Injury, operations,  
general anesthesia

# CAUSES OF PRIMARY IMMUNODEFICIENCY

Contoh Penyakit	Kelainan/kerusakan yang disebabkan	Dampak klinis
Defisiensi imunitas kombinasi (Severe Combined Immunodeficiency/SCID)	Penurunan jumlah sel T, Sel B, sel NK, dan atau antibody	Rentan terhadap infeksi virus, fungi, dan bakteri karena kecacatan pada system kekebalan selular dan humoral
X-linked agammaglobulinemia	Kegagalan maturase sel B di sumsum tulang belakang	Penurunan atau sama sekali tidak ada produksi sel B dan antibody
Sindrom DiGeorge	Ketidak sempurnaan perkembangan organ timus dan kegagalan maturase sel T	Rentan terhadap infeksi virus dan fungi karena kegagalan sistem imunitas humoral
Sindrom Wiskott-Aldrich	Cacat fungsi trombosit, sel T, dan kekurangan antibody (terutama IgA)	Rentan terhadap eczema atopik dan infeksi yang mudah kambuh
Hyper-IgM syndrome	Cacat pada sel B sehingga tidak dapat melakukan pergantian kelas antibody (immunoglobulin)	Kadar IgM didalam tubuh menjadi berlebih dan kekurangan IgA, IgG, dan IgE. Hal ini menyebabkan erring terjadinya infeksi berulang

# WISKOTT-ALDRICH SYNDROME

[ECZEMA-THROMBOCYTOPENIA-IMMUNODEFICIENCY SYNDROME]

ATOPIC DERMATITIS



PROBLEM with IMMUNE SYSTEM

↓  
REPEATED  
INFECTIONS

MICROTHROMBOCYTOPENIA

- VERY FEW PLATELETS

SMALL →



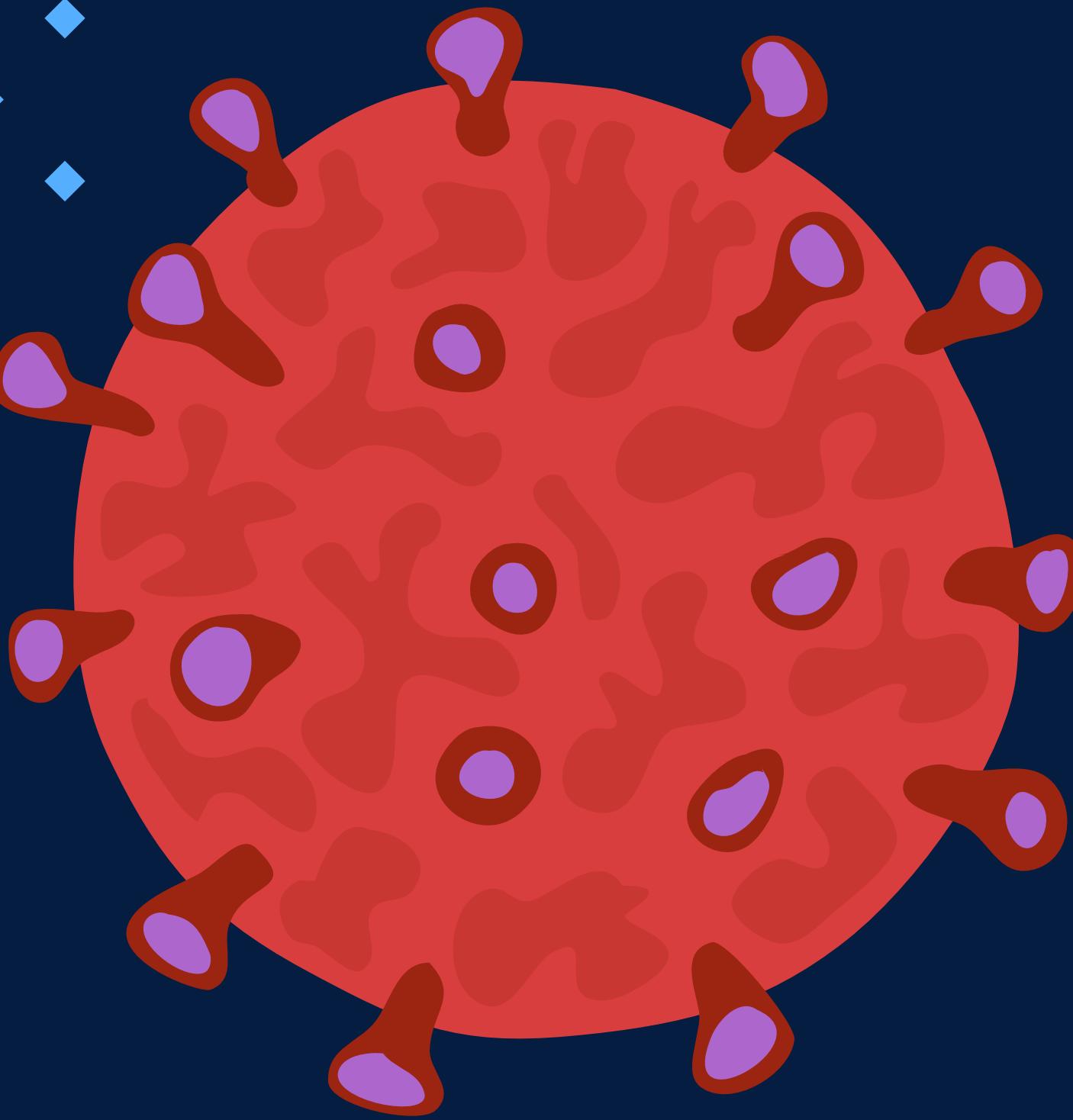


# ATAKSIA TELANGIEKSIA

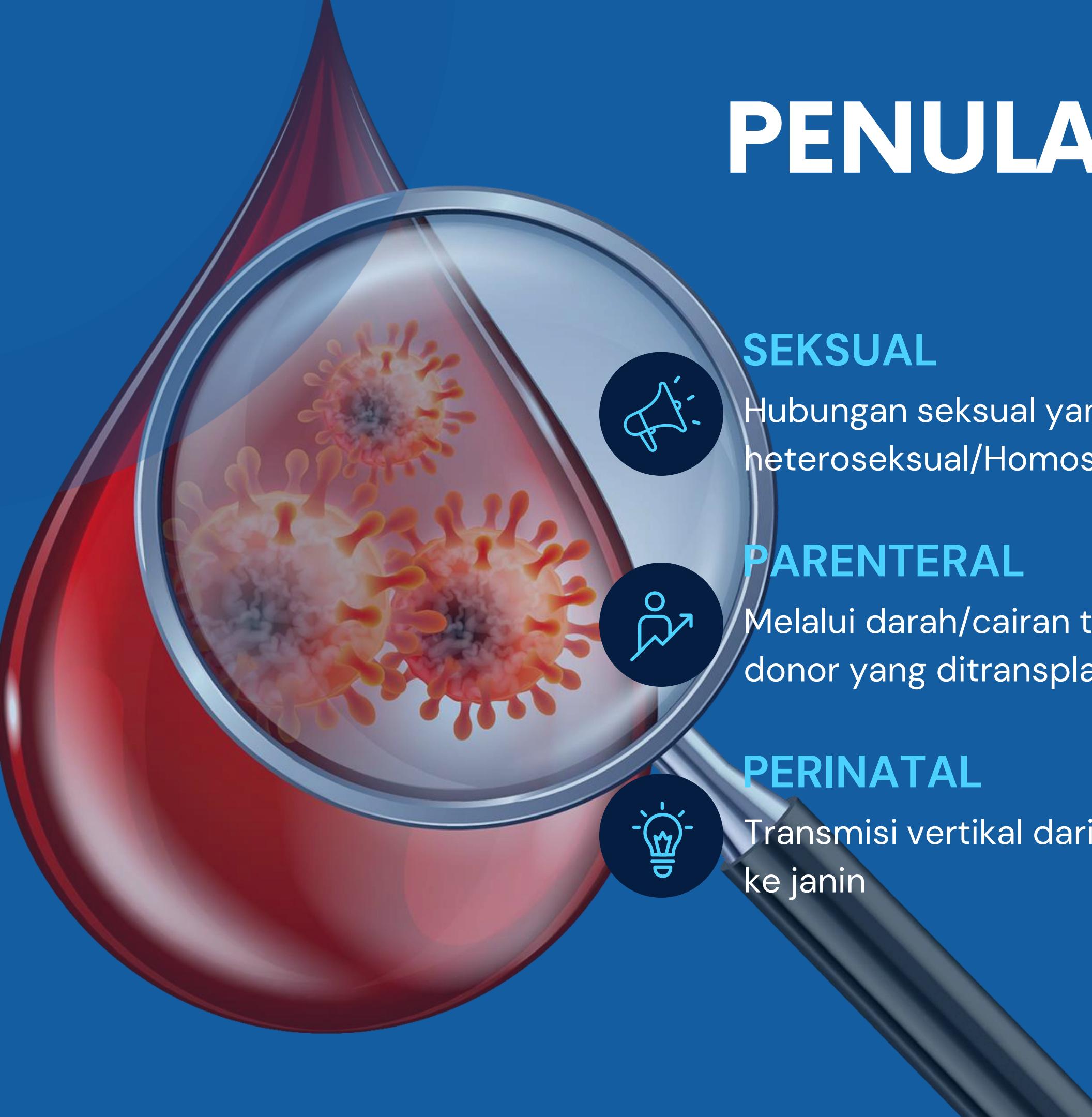
Penyakit autosomal resesif mengenai syaraf, endokrin,d an sistem vaskuler  
Ciri klinisnya berupa gerakan otot yang tidak terkoordinasi dan dilatasi pembulih sarah kecil terlihat di sklera mata, limfopenia, penurunan Ig A, Ig E, dan kadang Ig G.

# AIDS

*Aquired Immunodeficiency Syndrome* adalah suatu penyakit retrovirus epidemik, menular yang disebabkan infeksi *Human Immunodeficiency Virus*, yang pada kasus berat bermanifestasi depresi berat imunitas selular.



# PENULARAN HIV



## SEKSUAL

Hubungan seksual yang tidak aman,  
heteroseksual/Homoseksual

## PARENTERAL

Melalui darah/cairan tubuh/semen/organ  
donor yang ditransplantasi

## PERINATAL

Transmisi vertikal dari ibu yang terinfeksi HIV  
ke janin

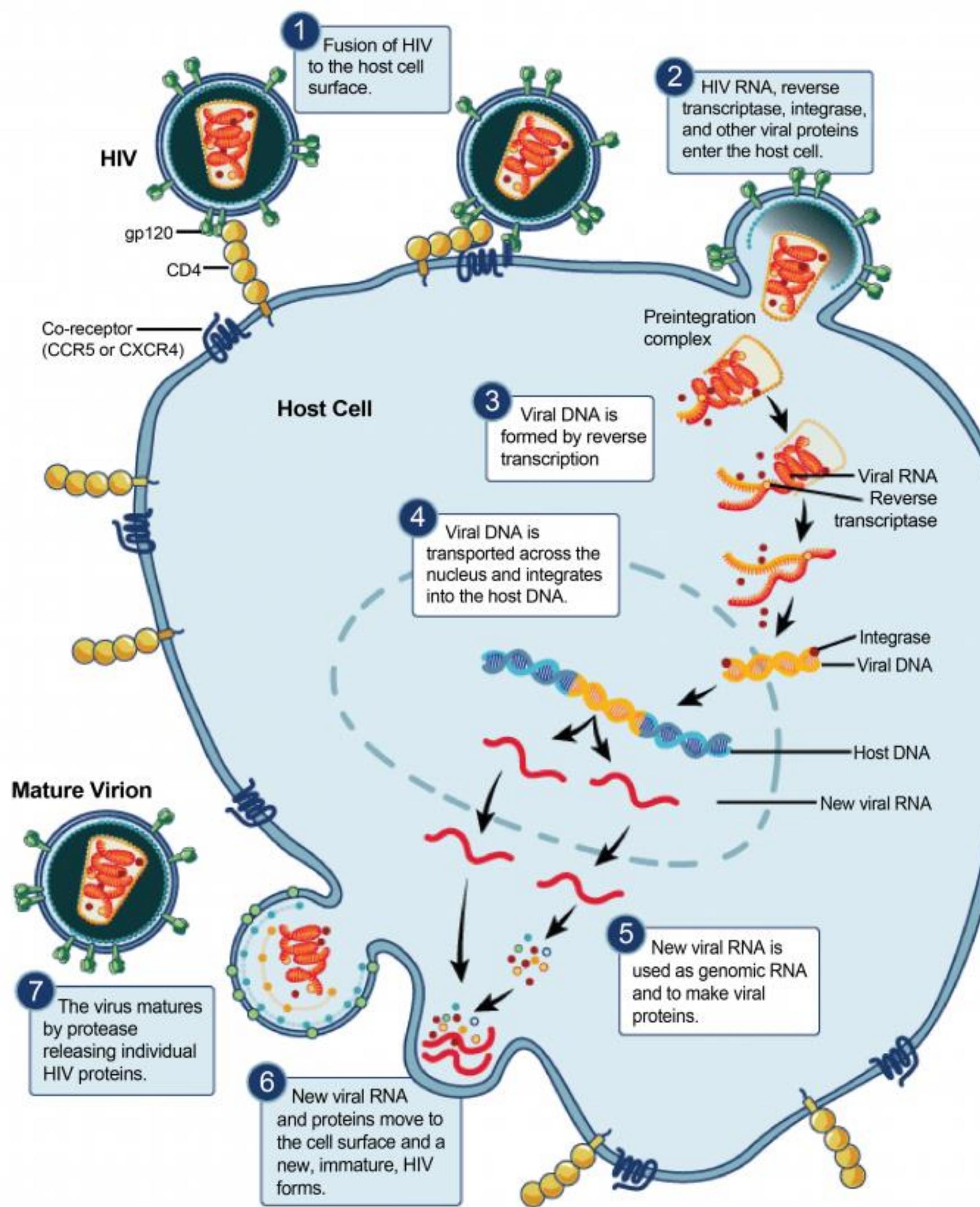
# PENULARAN HIV PADA KEHAMILAN

intrauterin

intrapartum

Pasca  
persalinan

- ✓ Faktor virus : makin tinggi titer virus , makin infeksius.
- ✓ Faktor Host (ibu hamil) : sistem kekebalan tubuh, nutrisi, anemia.
- ✓ Faktor Obstetrik : lama dan cara persalinan.
- ✓ Faktor bayi : Menyusui



## PATHWAY PATOFISIOLOGI HIV/AIDS

Transmisi HIV ke dalam tubuh melalui darah

Pengikatan gp120 HIV dengan reseptor membran T Helper + CD4

Fusi / peleburan membran virus dengan membran sel T Helper + CD4

**Enzim reverse transcriptase**

RNA HIV → cDNA

**Enzim integrase**

cDNA masuk ke inti sel T Helper

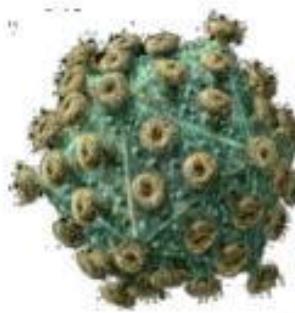
Transkripsi mRNA dan translasi menghasilkan protein struktural virus

**Enzim protease**

Merangkai RNA virus dengan protein-protein yang baru dibentuk,

Terbentuk virus - virus HIV yang baru dalam tubuh

# TAHAPAN PERKEMBANGAN HIV MENJADI AIDS



**Infeksi primer:**  
Virus masuk ke tubuh

**Masa Jendela** (3 – 6 bulan): Saat Virus mulai membangun kekuatan untuk menyerang. Pada tahapan ini, virus tidak terdeteksi namun bisa menularkan

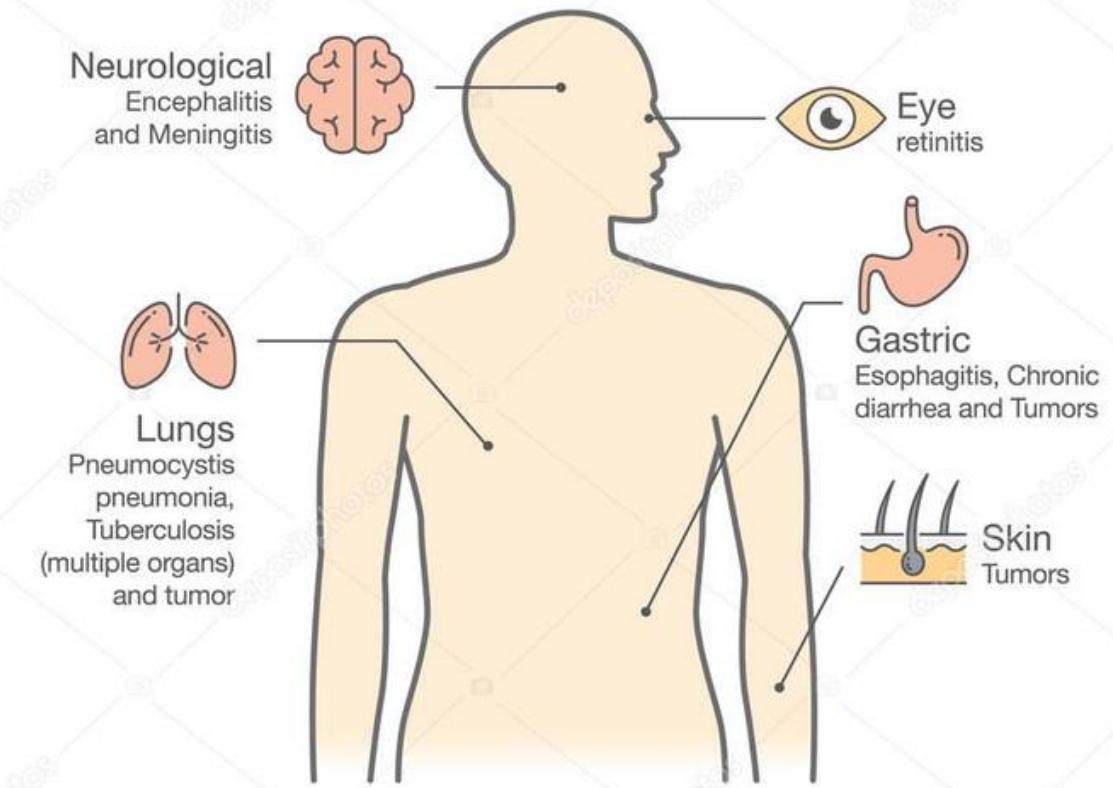
**Tahapan tanpa gejala** (HIV +): 3 – 10 tahun. Pada tahapan ini, virus sudah mulai berkembang, tapi penderita masih bisa beraktifitas normal seperti biasa. Dan **tidak ada tanda-tanda** tertentu pada penderita

**Tahapan gejala awal:** pada tahap ini, mulai muncul penyakit-penyakit ringan pada penderita seperti: Diare, demam, berat badan turun, infeksi pada mulut yang berkepanjangan

**Tahapan gejala lanjutan** (AIDS): mulai munculnya penyakit-penyakit yang lebih berat, seperti: PCP, CMV, TB, Candida, Sarkoma kaposi, dementia complex, turunnya berat badan yang drastis.



Main symptoms of AIDS





# DEFISIENSI AUTOIMUNE

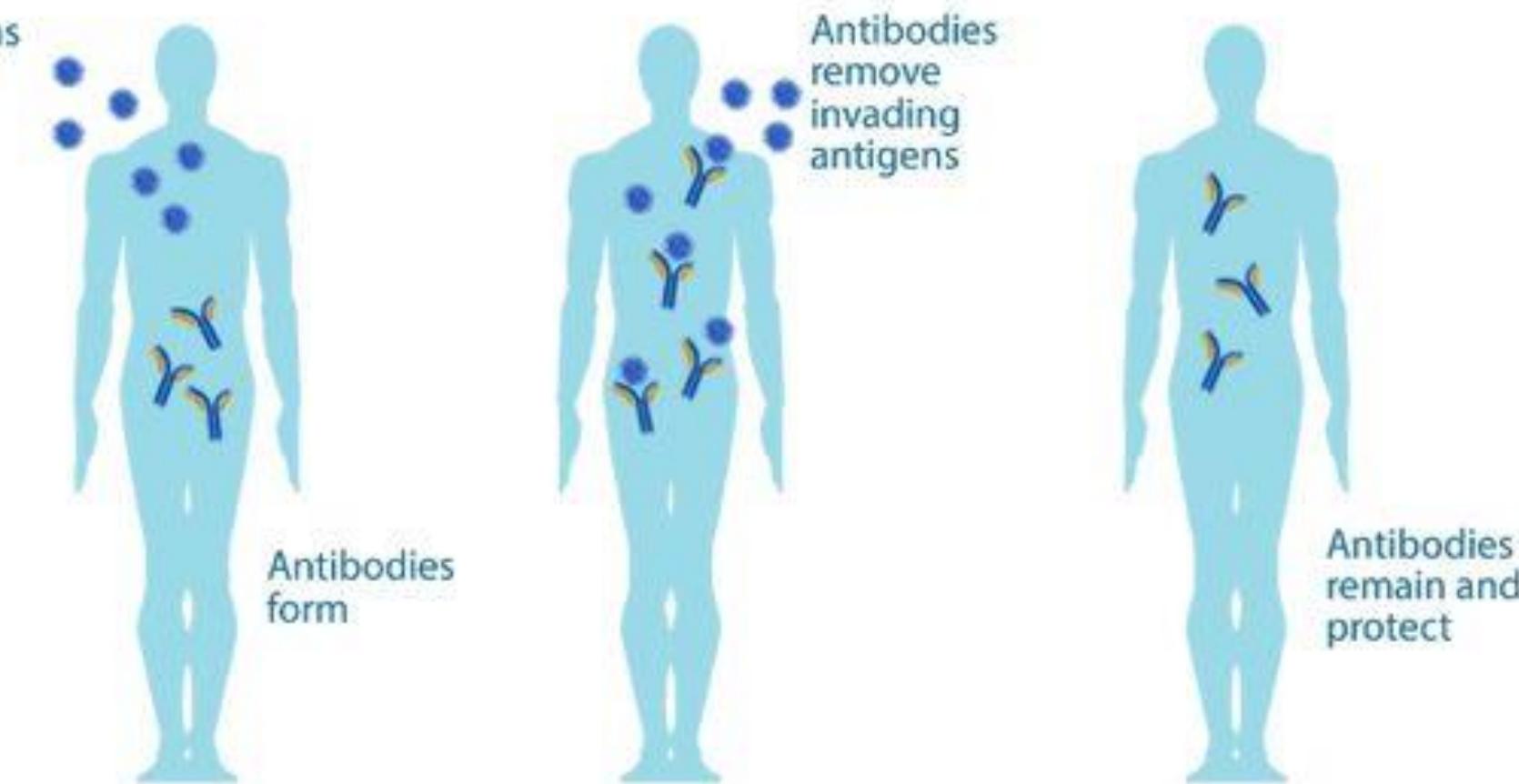
## AUTOIMUNITAS

Merupakan respon imun terhadap antigen jaringan sendiri yang disebabkan oleh kegagalan mekanisme mempertahankan toleransi sel B, sel T, atau keduanya.

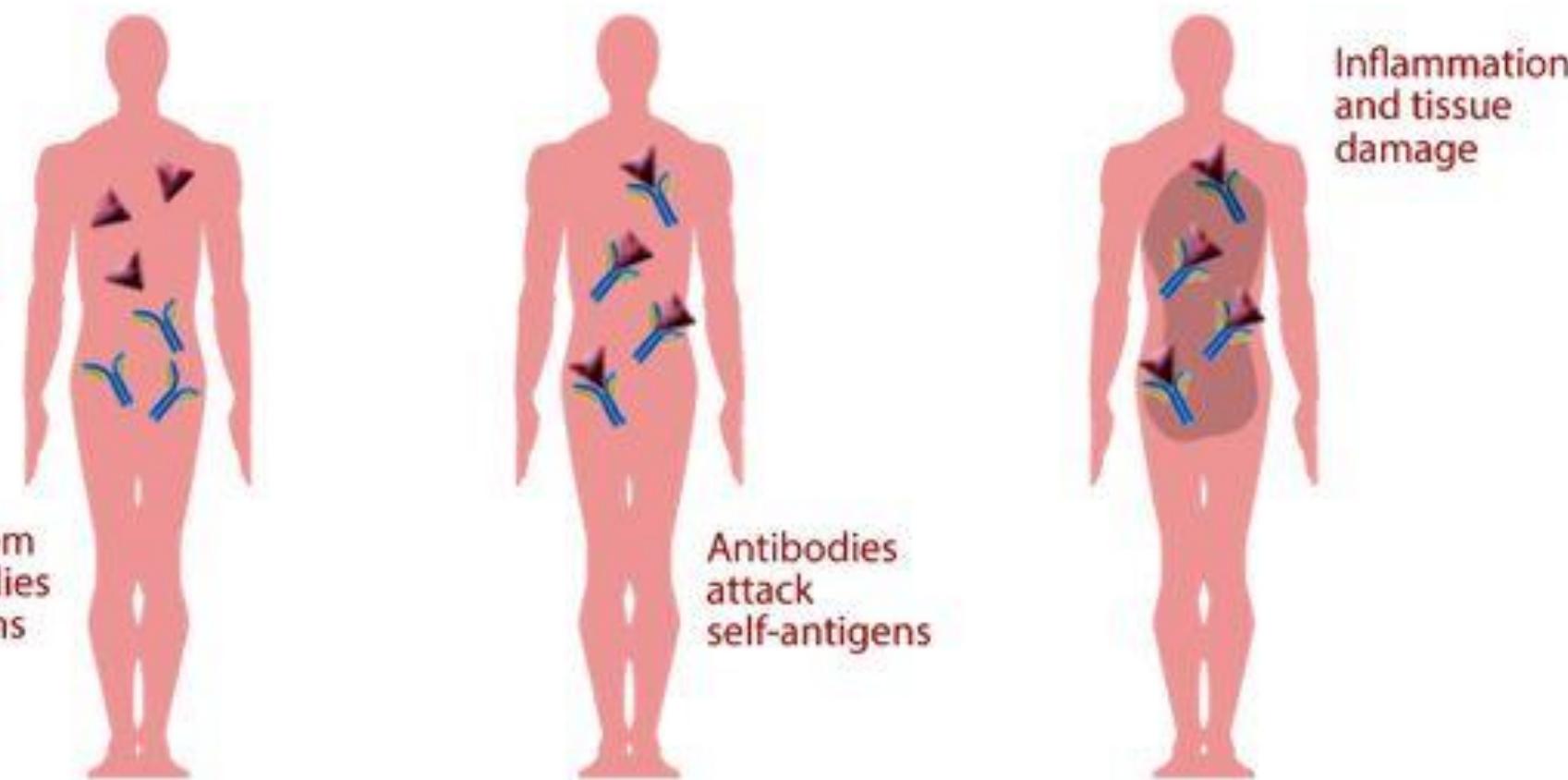
Kegagalan pada toleransi imunitas sendiri

Penyakit autoimun adalah kerusakan jaringan atau gangguan fungsi fisiologis yang ditimbulkan oleh respon autoimun

## NORMAL IMMUNE RESPONSE



## AUTOIMMUNE DISEASE



# ETIOLOGI AUTOIMUN



1

Reaksi silang  
dengan antigen  
bakteri



2

Rangsangan  
molekul poliklonal



3

Kegagalan  
utoregulasi

# AUTOIMMUNE DISEASES

## Brain

Multiple Sclerosis  
Guillain-Barre Syndrome  
Autism



## Thyroid

Thyroiditis  
Hashimoto's Disease  
Graves' Disease



## Blood

Leukemia  
Lupus Erythematosus  
Hemolytic Dysglycemia



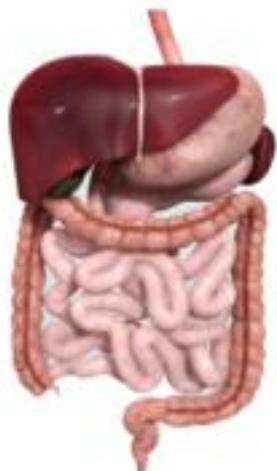
## Bones

Rheumatoid Arthritis  
Ankylosing Spondylitis  
Polymyalgia Rheumatica



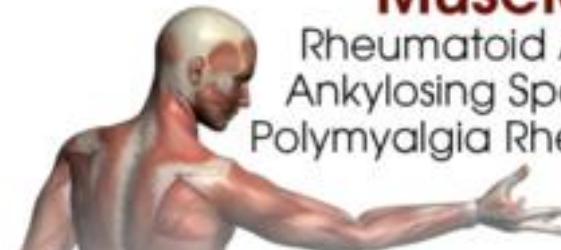
## GI Tract

Celiac's Disease  
Crohn's Disease  
Ulcerative Colitis  
Diabetes Type I



## Muscles

Rheumatoid Arthritis  
Ankylosing Spondylitis  
Polymyalgia Rheumatica



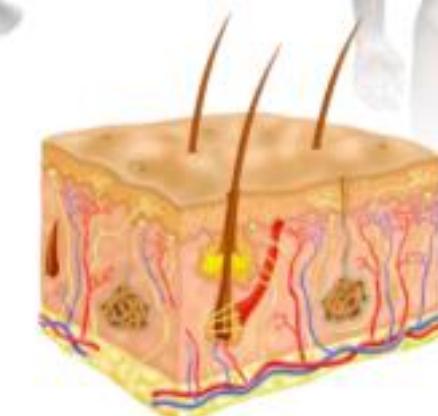
## Nerves

Peripheral Neuropathy  
Diabetic Neuropathy



## Skin

Psoriasis  
Vitiligo  
Eczema  
Scleroderma

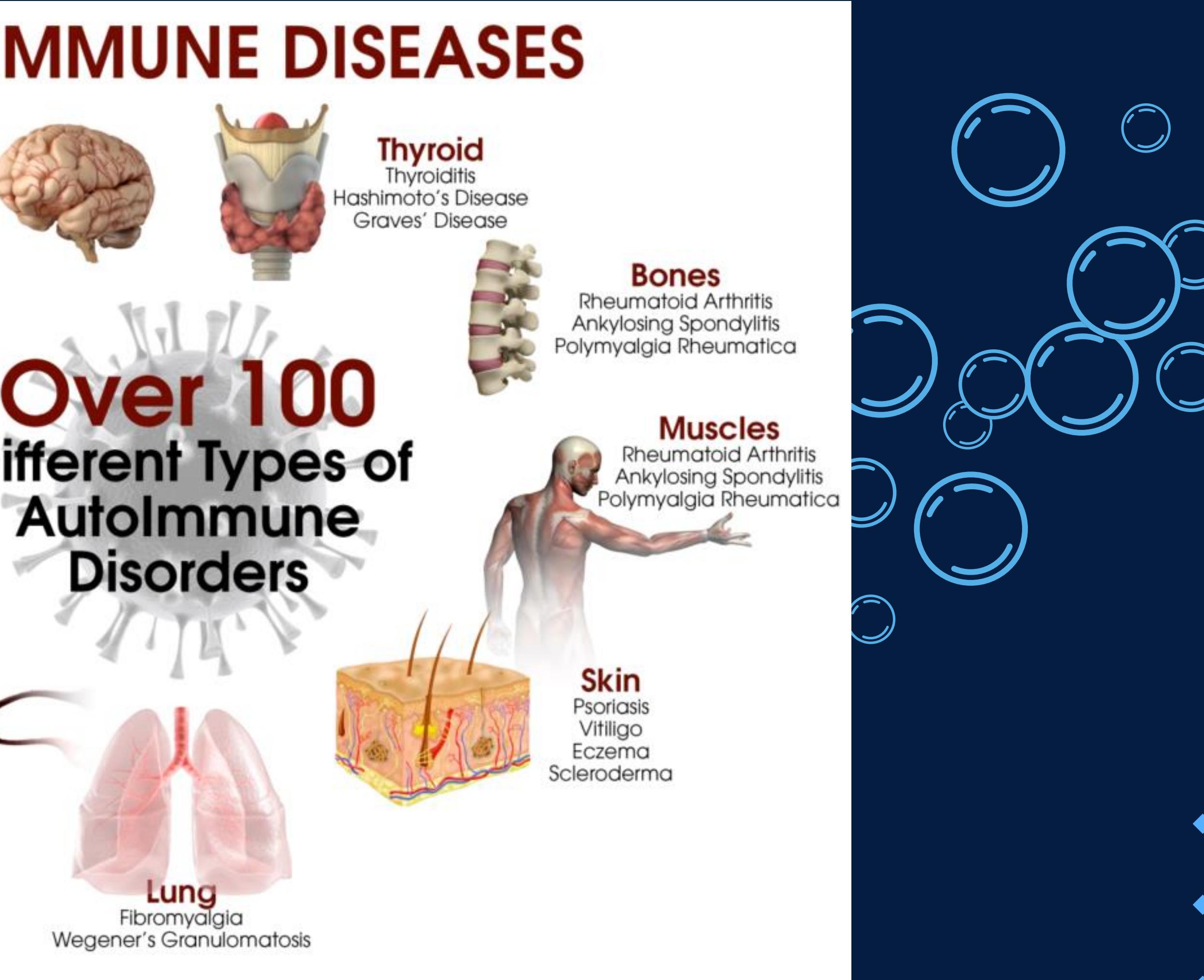


## Lung

Fibromyalgia  
Wegener's Granulomatosis



Over 100  
Different Types of  
Autoimmune  
Disorders



# DM TIPE 1



## Pathogenesis of Type I DM

### Other Autoimmune disorders:

- PS Glomerulonephritis
- Graves, Hashimoto thyroiditis.
- Rheumatic heart disease
- SLE, Collagen vascular disease
- Rheumatoid arthritis.

### Antibodies:

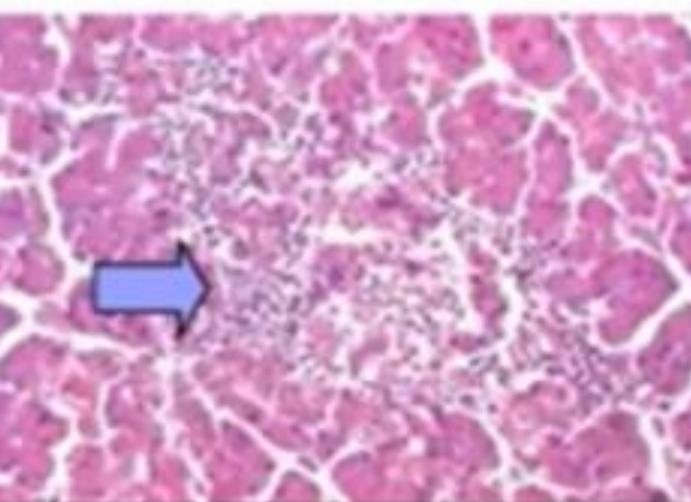
Islet cell Ab - **ICA**

Insulin Auto Ab - **IAA**

Glut. Acid Decarb - **GAD65**

Environment  
Viral infec..?

Genetic  
HLA-DR3/4



Insulin deficiency

$\beta$  cell  
Destruction

Autoimmune Insulitis  
Ab to  $\beta$  cells/insulin

Secondary DM  
Inflammation,  
Tumor,  
Infection  
Trauma  
Pancreatitis



# Pathophysiology of Diabetes Mellitus Type 2

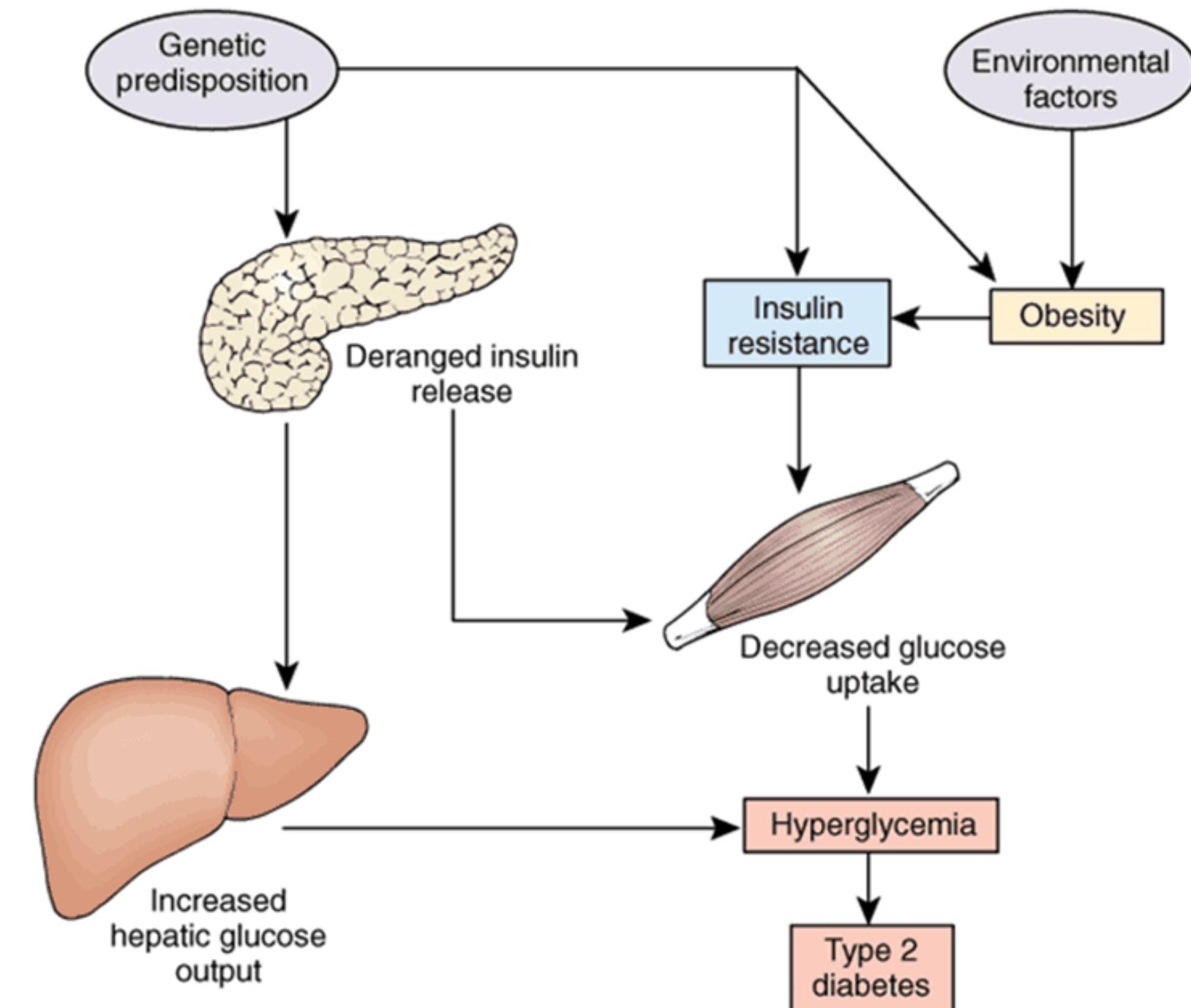
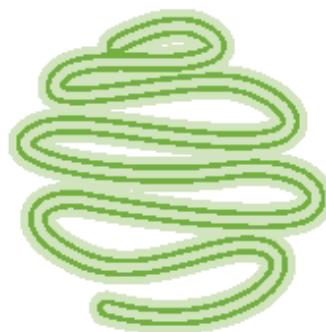


Image via: [proprofs.com](https://www.proprofs.com)

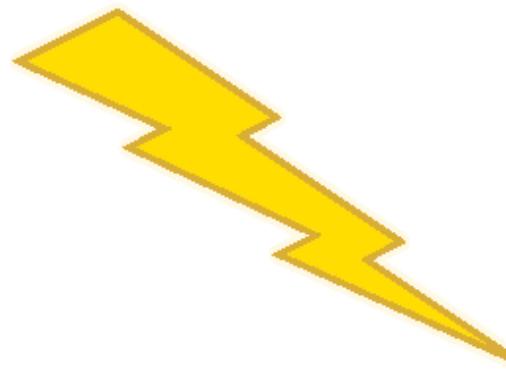
# SLE

## 1. Genes



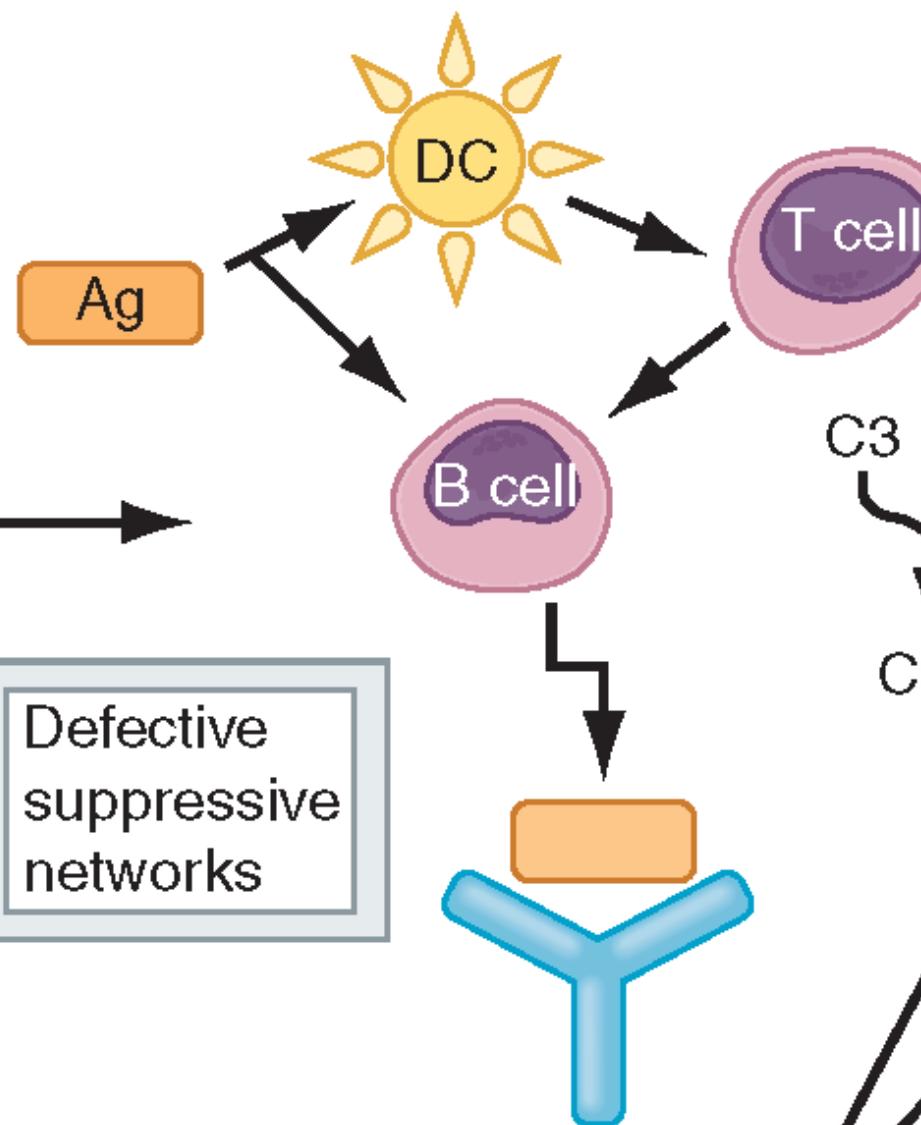
C1q,C2,C4  
HLA-D2,3,8  
MBL  
FcR 2A,3A,2B  
IL-10  
MCP-1  
PTPN22

## Environment



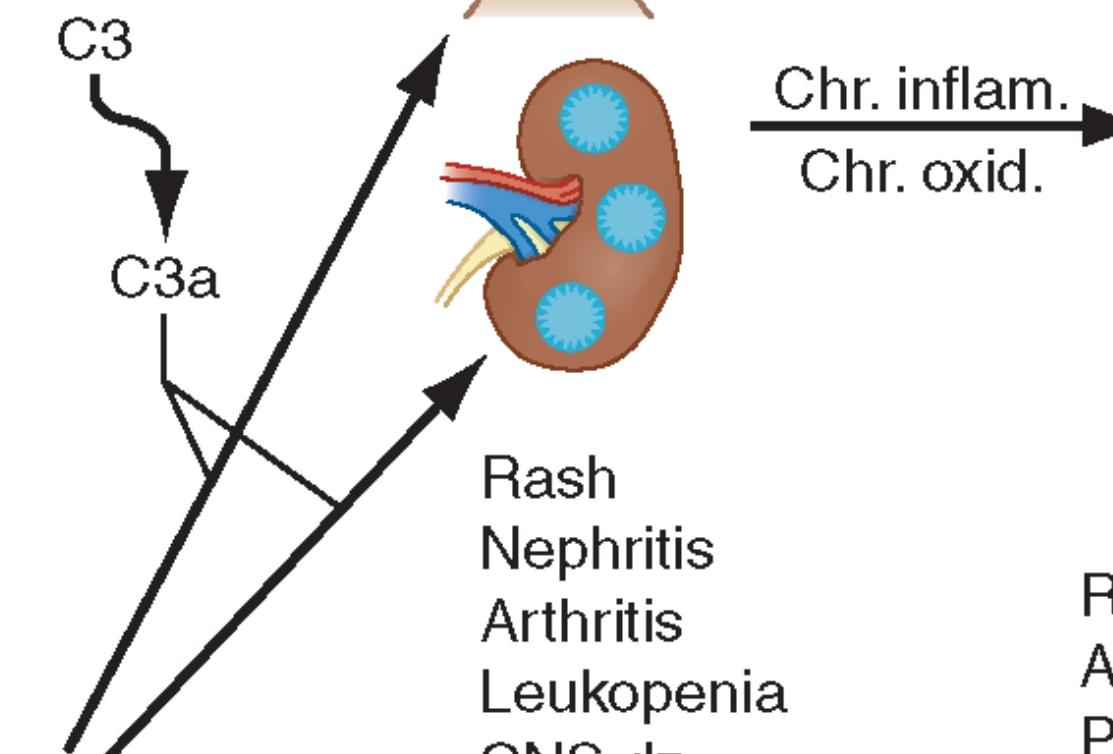
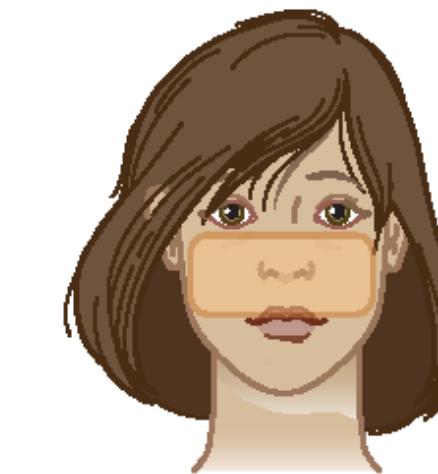
UV light  
Gender  
?Infection  
?EBV  
Others

## 2. Abnormal Immune Response

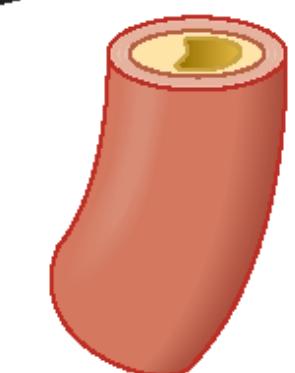
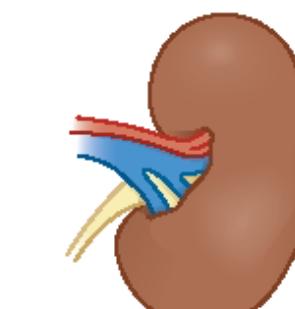


## 3. Autoantibodies Immune Complexes

## 4. Inflammation



## 5. Damage



Renal Failure  
Atherosclerosis  
Pulm fibrosis  
Stroke  
Damage from Rx  
Etc.

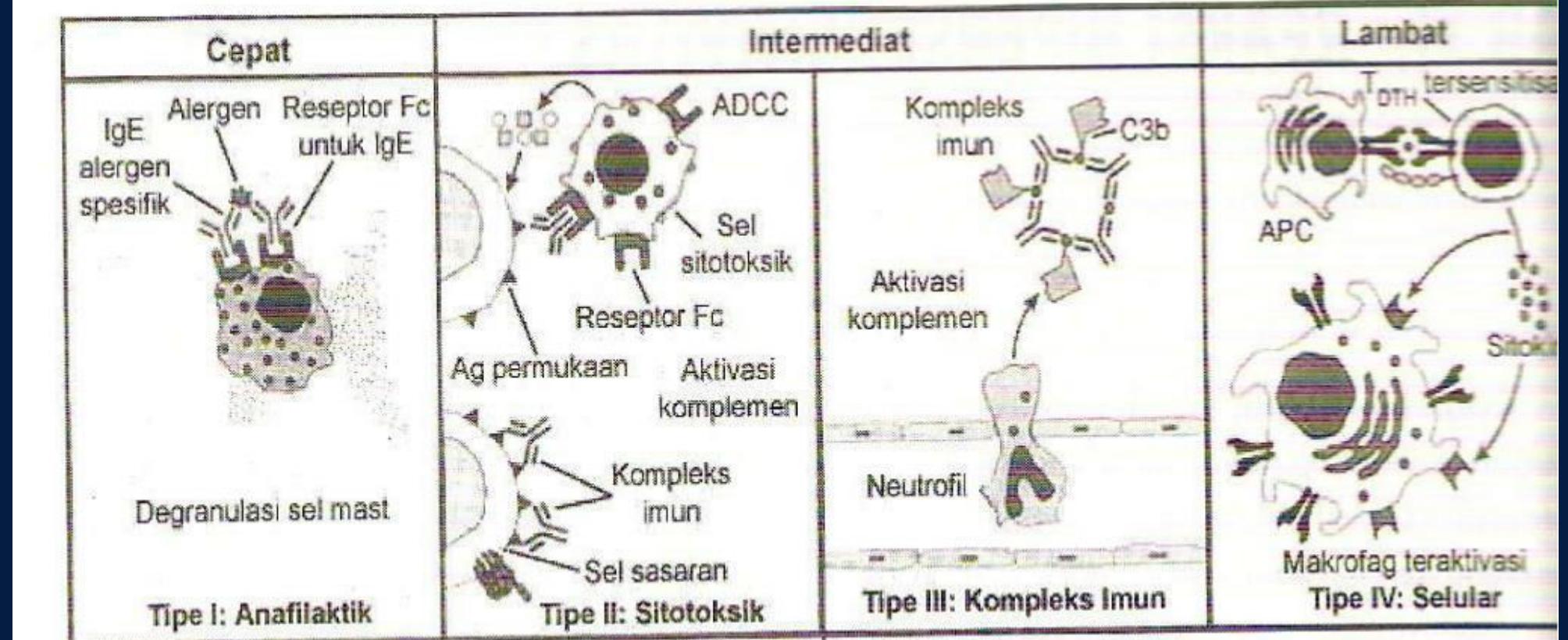
# HIPERSENSITIFITAS

Hipersensitivitas :  
Reaksi imun yang patologik, terjadi akibat respons imun yang berlebihan menimbulkan kerusakan jaringan tubuh.

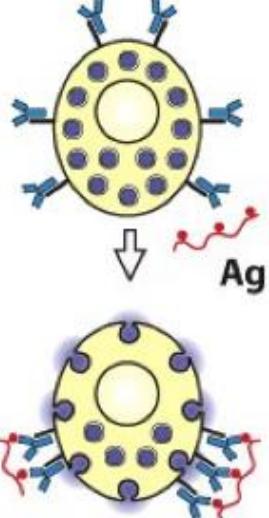
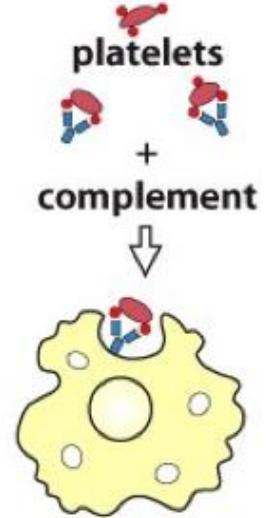
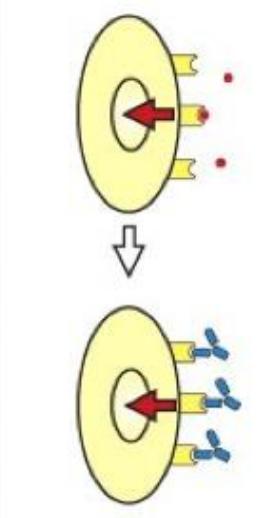
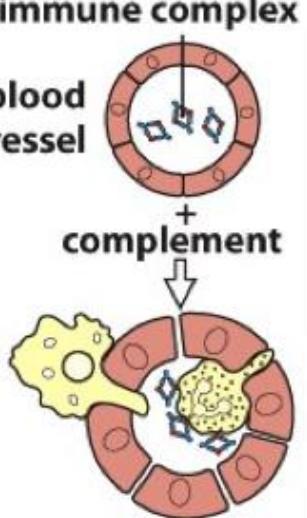
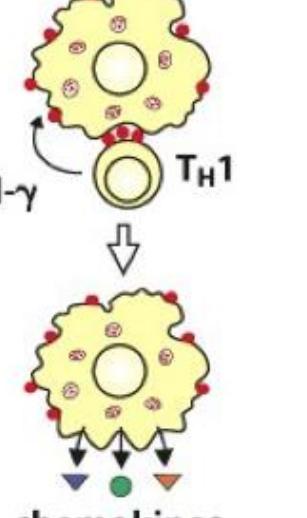
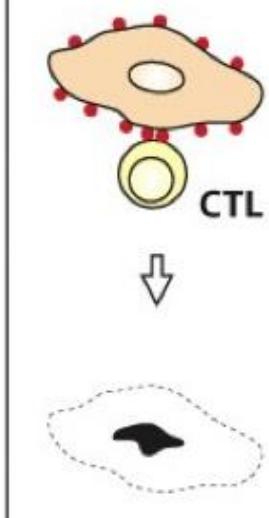
Gell dan Coombs :

- Membagi dalam tipe I, II, III & IV berdasarkan kecepatan dan mekanisme imun yang terjadi.

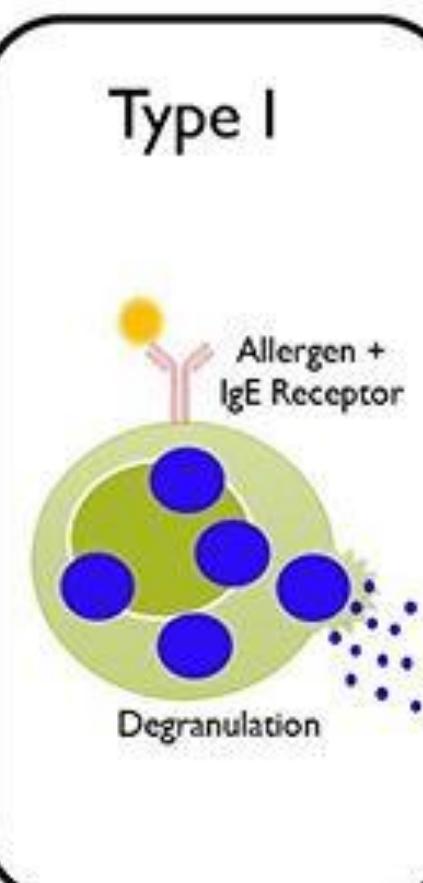
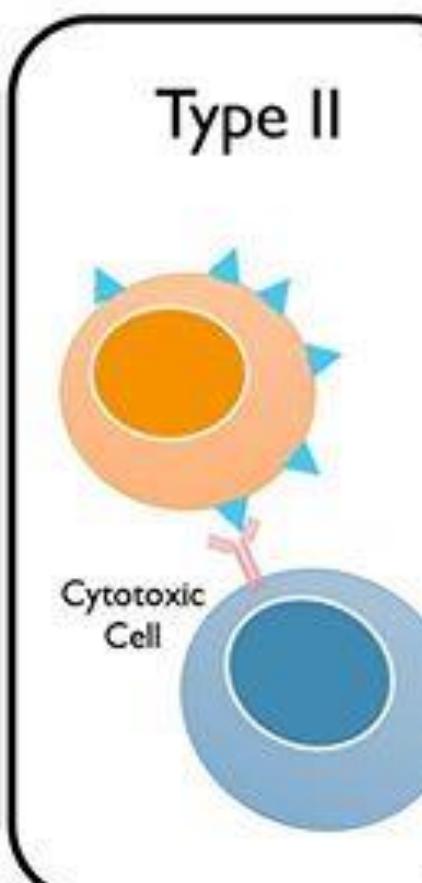
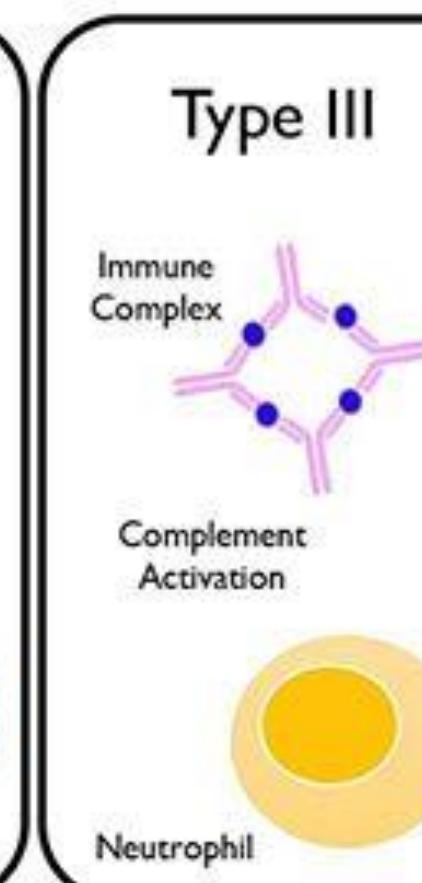
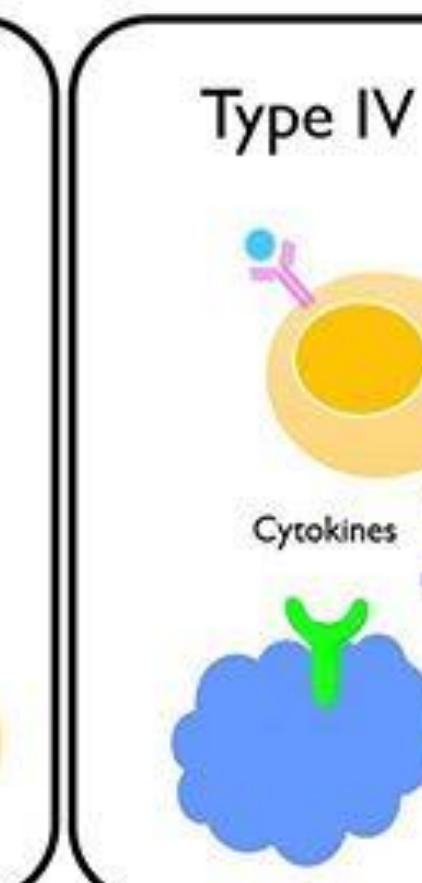
## I. PEMBAGIAN REAKSI HIPERSENSITIFITAS MENURUT WAKTU



Gambar 115. Berbagai jenis respons hipersensitivitas: cepat, intermediat dan lambat

	Type I	Type II	Type III	Type IV			
Immune reactant	IgE	IgG		IgG	$T_{H1}$ cells	$T_{H2}$ cells	CTL
Antigen	Soluble antigen	Cell- or matrix-associated antigen	Cell-surface receptor	Soluble antigen	Soluble antigen	Soluble antigen	Cell-associated antigen
Effector mechanism	Mast-cell activation	Complement, $FcR^+$ cells (phagocytes, NK cells)	Antibody alters signaling	Complement, phagocytes	Macrophage activation	$IgE$ production, eosinophil activation, mastocytosis	Cytotoxicity
	 						
Example of hypersensitivity reaction	Allergic rhinitis, allergic asthma, atopic eczema, systemic anaphylaxis, some drug allergies	Some drug allergies (e.g. penicillin)	Chronic urticaria (antibody against $Fc\epsilon RI$ alpha chain)	Serum sickness, Arthus reaction	Allergic contact dermatitis, tuberculin reaction	Chronic asthma, chronic allergic rhinitis	Graft rejection, allergic contact dermatitis to poison ivy

# Types of Hypersensitivity Reactions

Type I	Type II	Type III	Type IV
			
IgE-Mediated	IgG or IgM Cytotoxic	Immune Complex-Mediated	T-Cell-Mediated
Within 1 Hour	Hours to Days	1-3 Weeks	Days to Weeks
Anaphylaxis	Hemolytic Anemia	Serum Sickness SLE	Rash SJS

THANK  
YOU!

