RHEUM SALAD

LUPUS
SCLERODERMA
REACTIVE ARTHRITIS

AMY RAMSAY, MD, FACEP

AUGUST 13, 2024

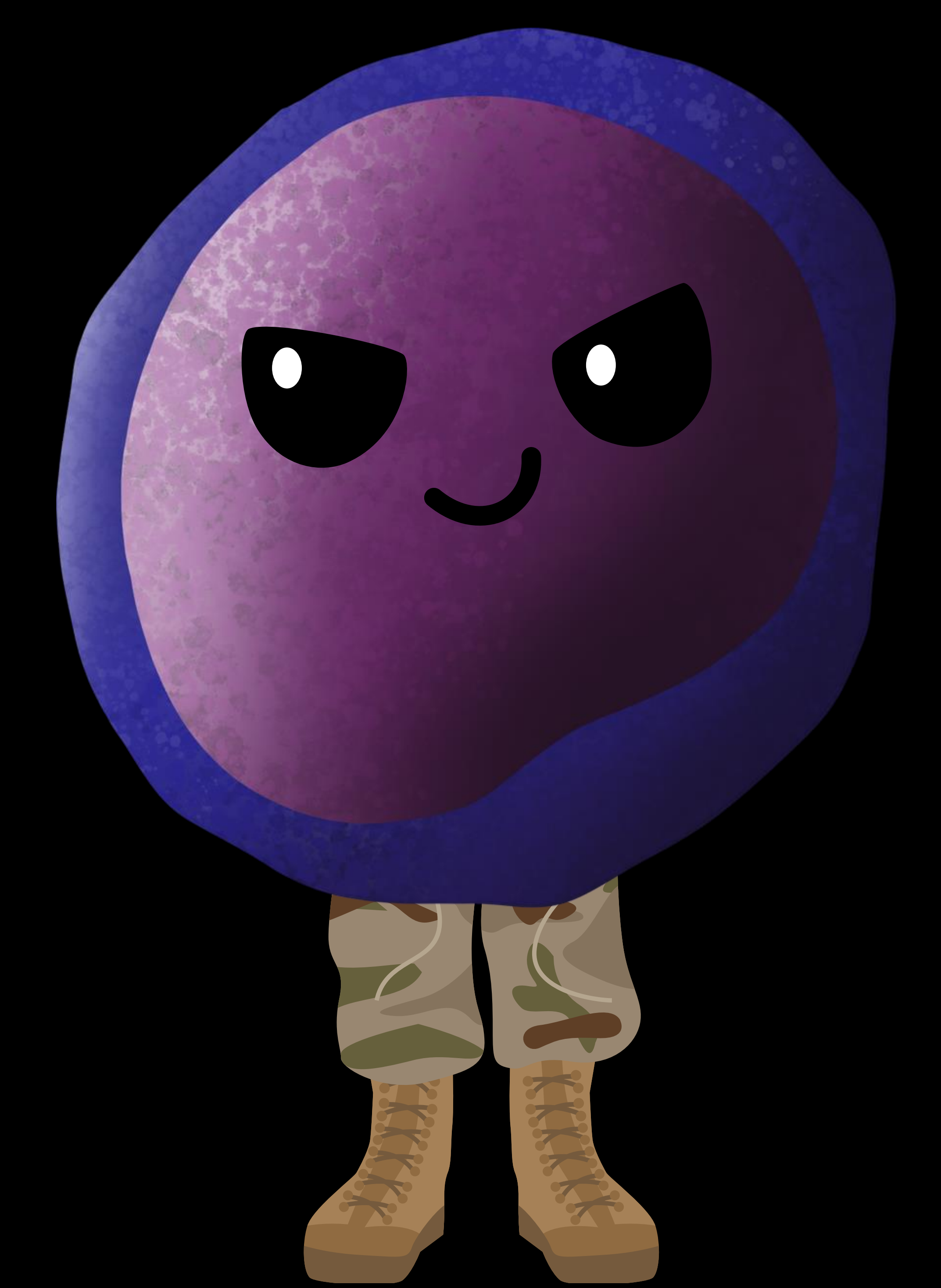
RHEUMATOLOGY
IMMUNOLOGY

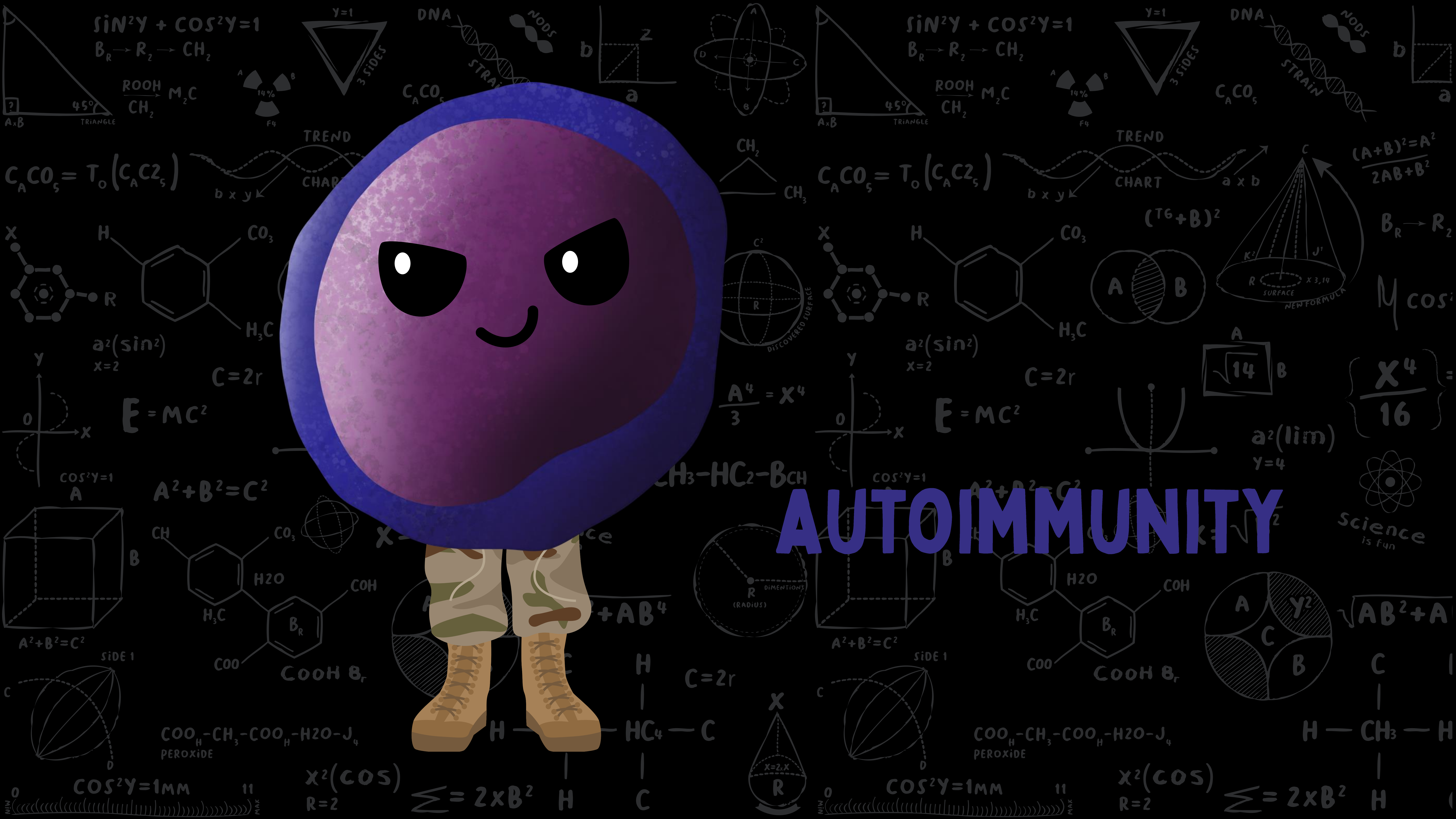
EMERGENCY
MEDICINE

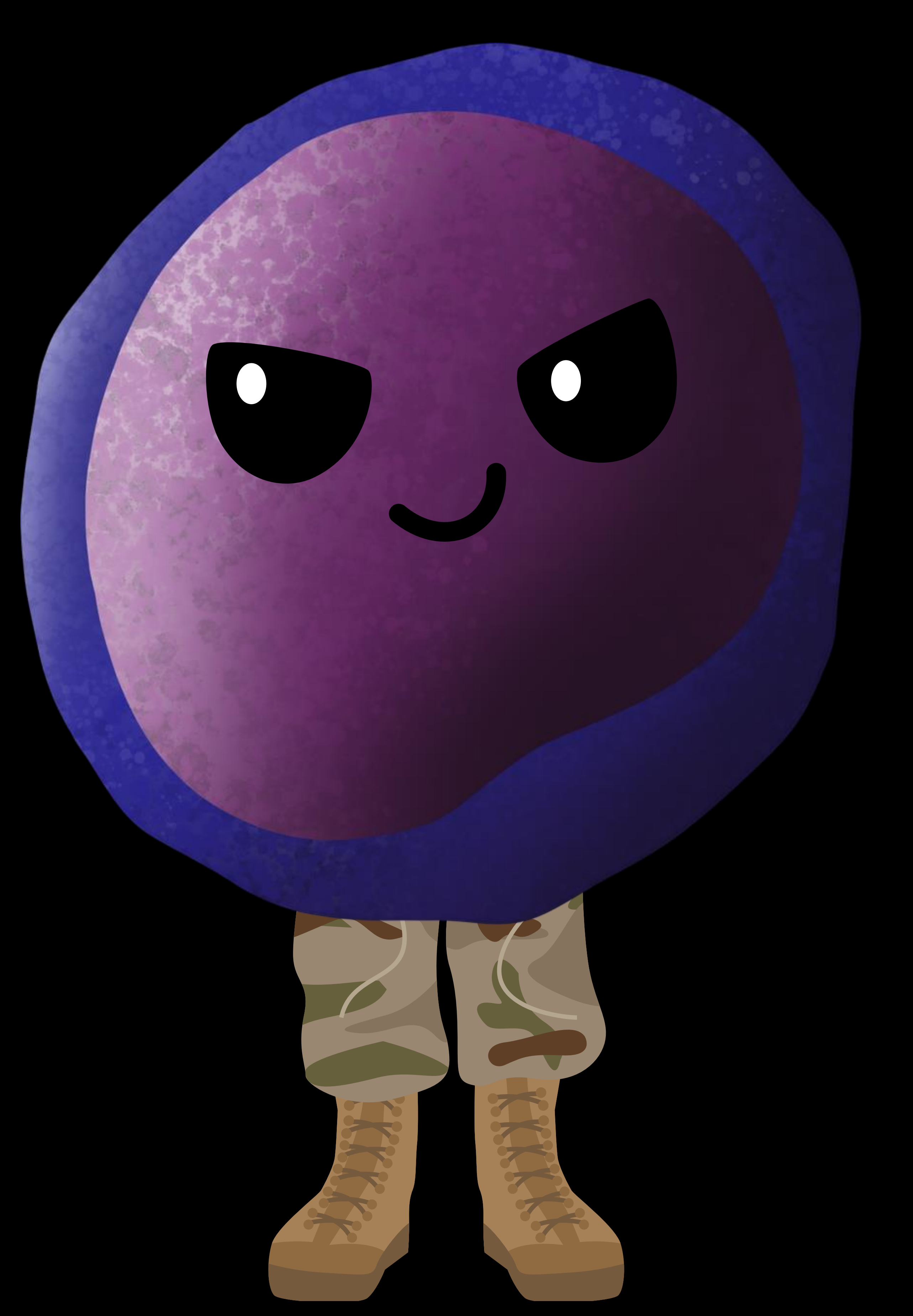
THERE IS SO MUCH
MORE TO READ!

PARIS 2024

PARIS 2024

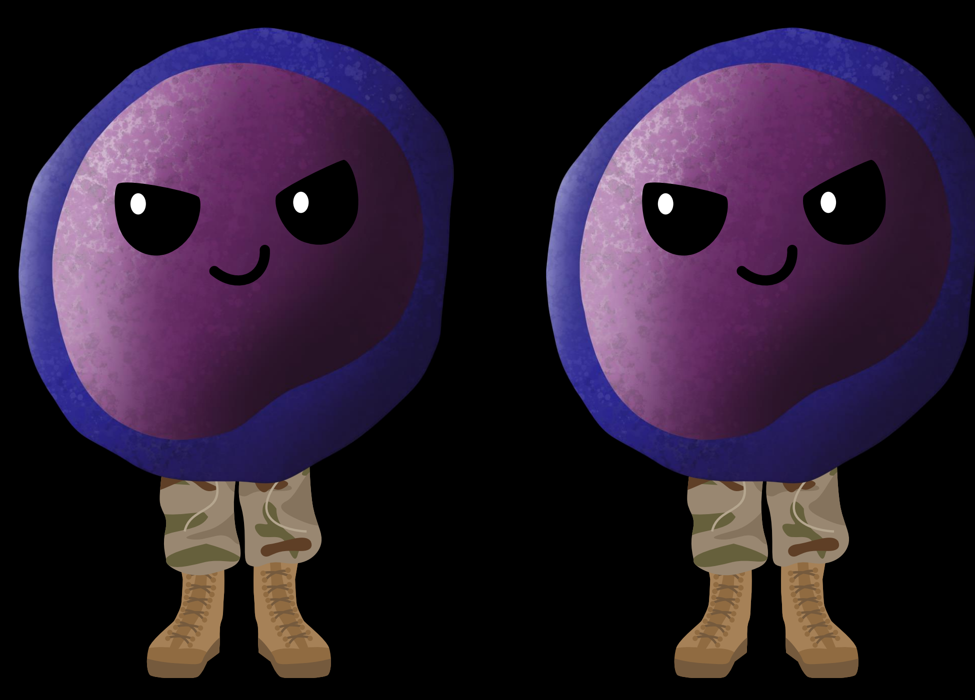



AUTOIMMUNITY



B LYMPHOCYTES AND T LYMPHOCYTES

B CELLS AND T CELLS



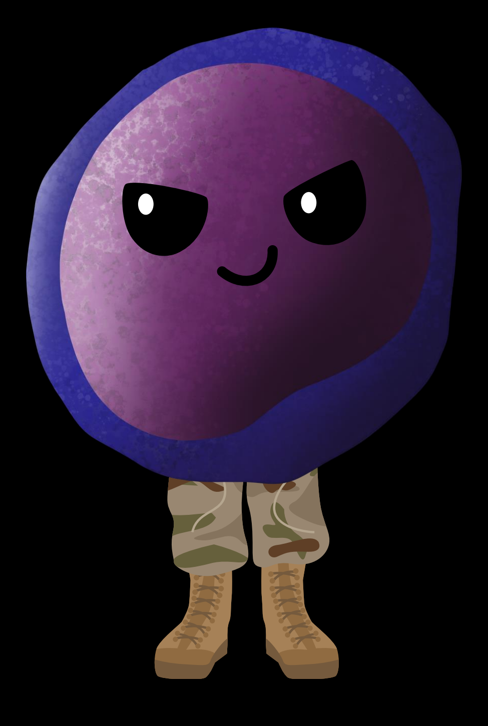
PLASMA CELLS

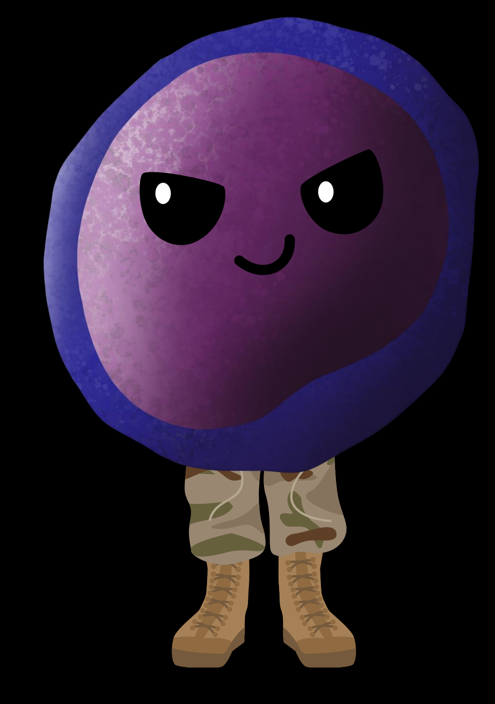
MAKE ANTIBODIES

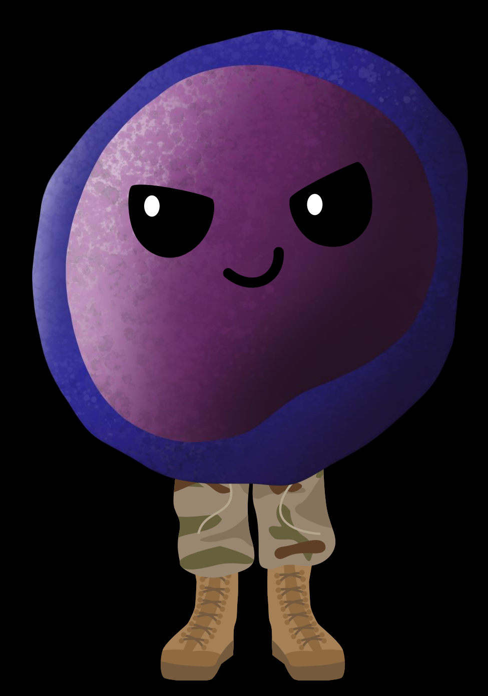
MEMORY B CELLS

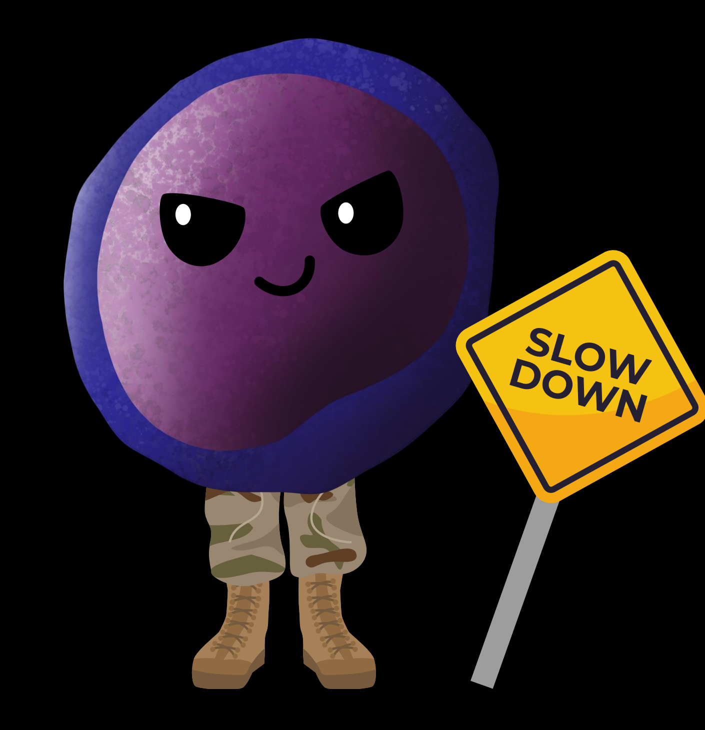
REMEMBER PATHOGENS FOR NEXT TIME

B CELLS AND T CELLS









PLASMA CELLS

MAKE ANTIBODIES

MEMORY B CELLS

REMEMBER PATHOGENS FOR NEXT TME

HELPER T CELLS

SOUND THE ALARM WITH CYTOKINES

KILLER T CELLS

CYTOTOXIC CELLS

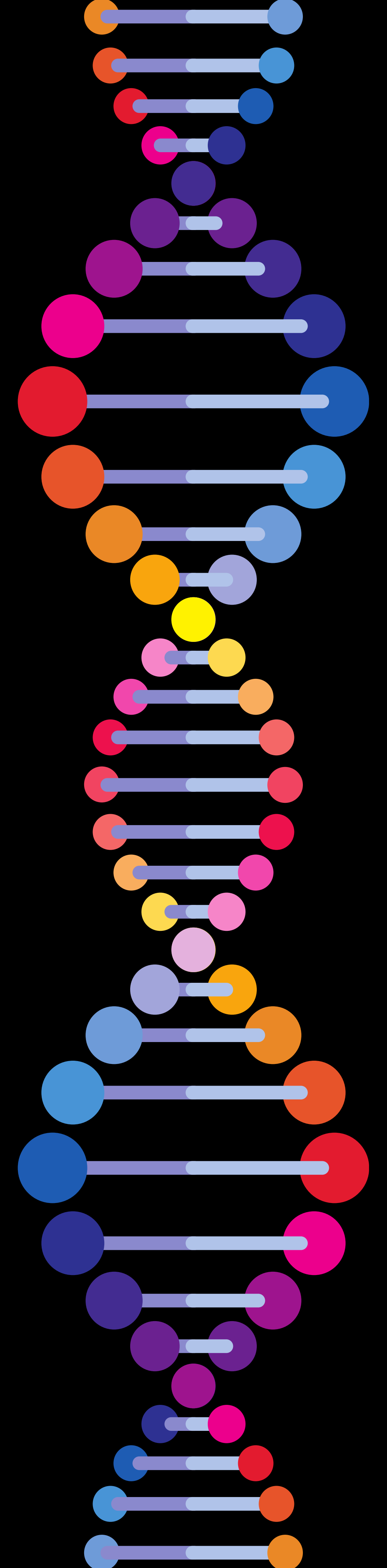
MEMORY T CELLS

HOLD ONTO ANTIGENS TO REMEMBER

REGULATORY T CELLS

PUT ON THE BRAKES

IMMUNE RESPONSE

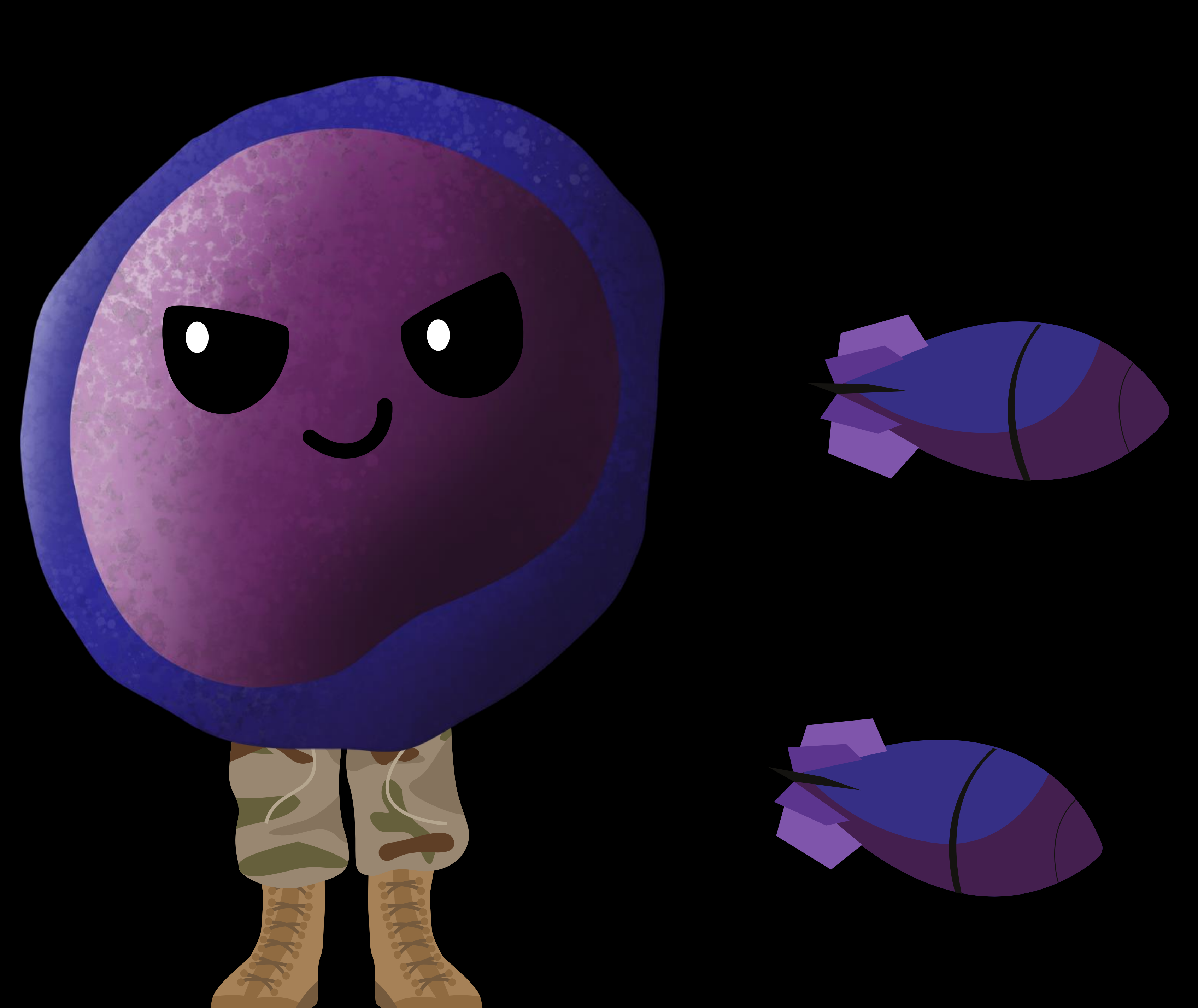


GENETIC PREDISPOSITION

TRIGGER

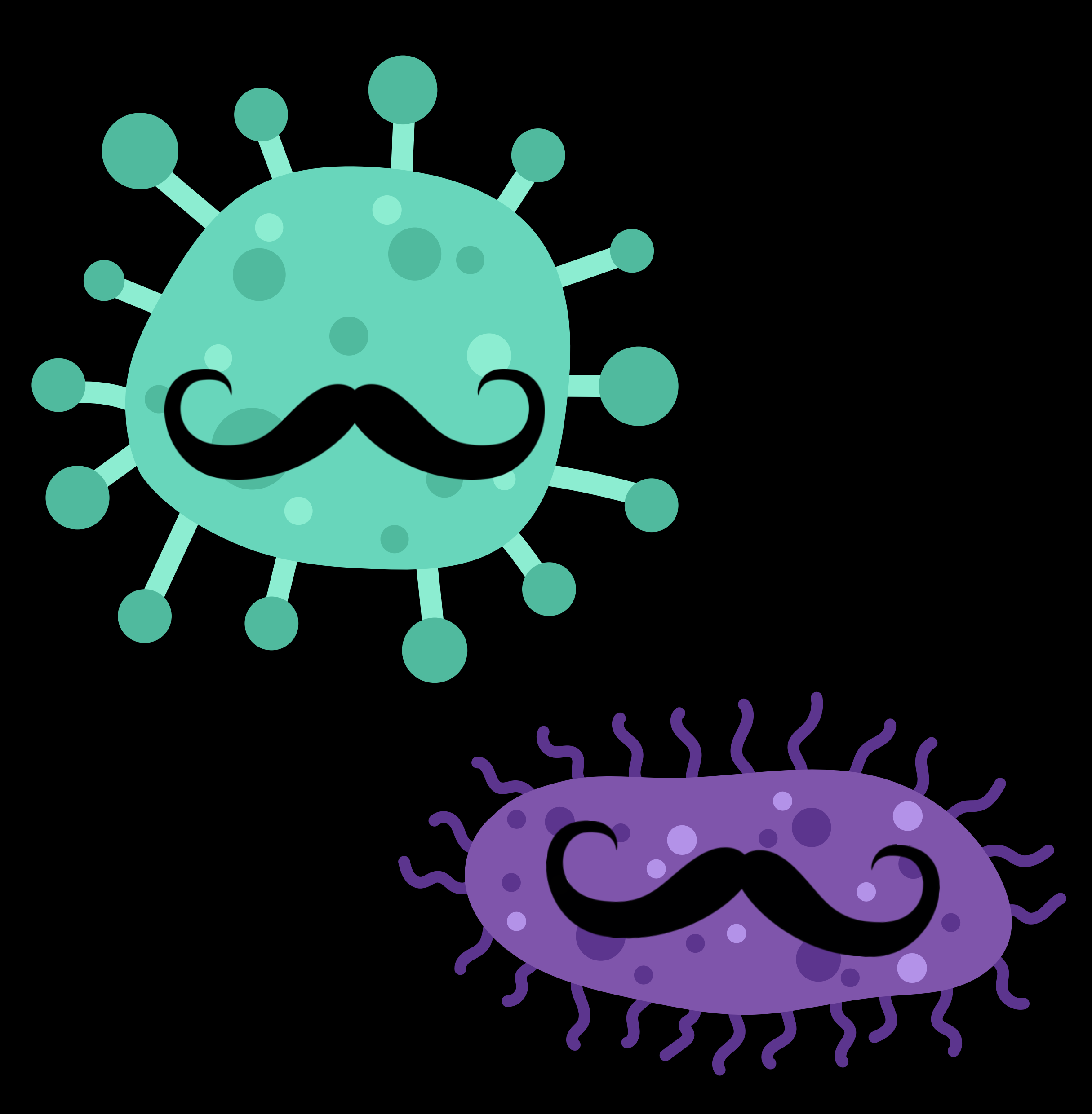
INFECTION OR OTHER
ENVIRONMENTAL TRIGGER

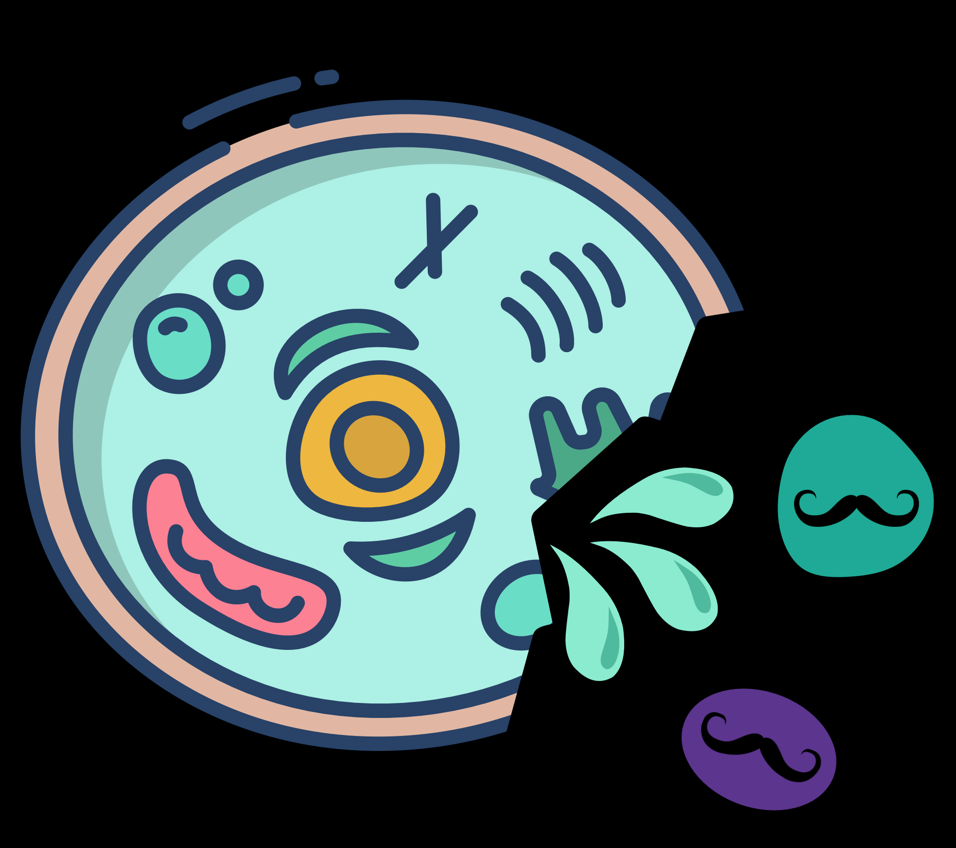
IMMUNE RESPONSE ATTACKS MICROBES



THOSE WITH GENETIC PREDISPOSITION

SOME SELF-PROTEINS LOOK EERILY SIMILAR TO FOREIGN ANTIGENS





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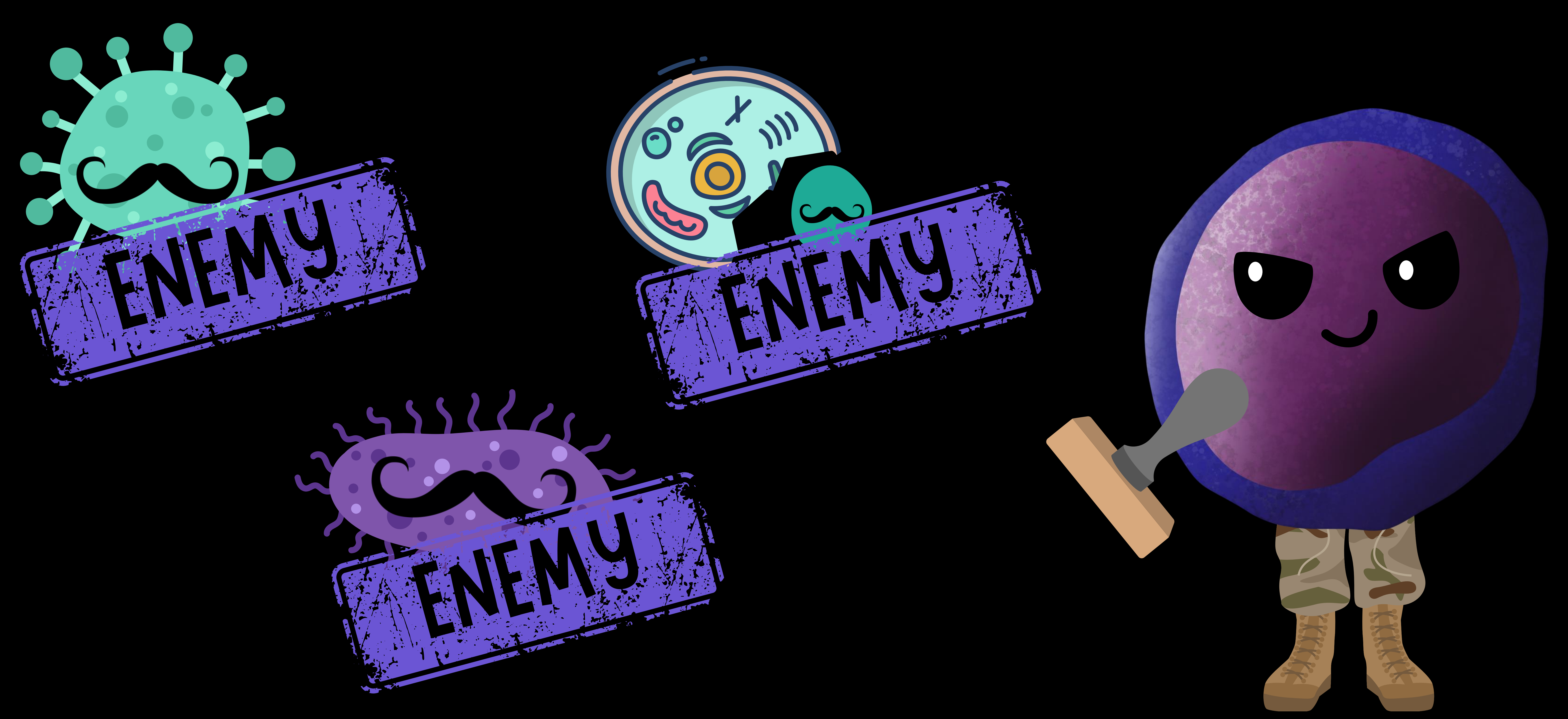


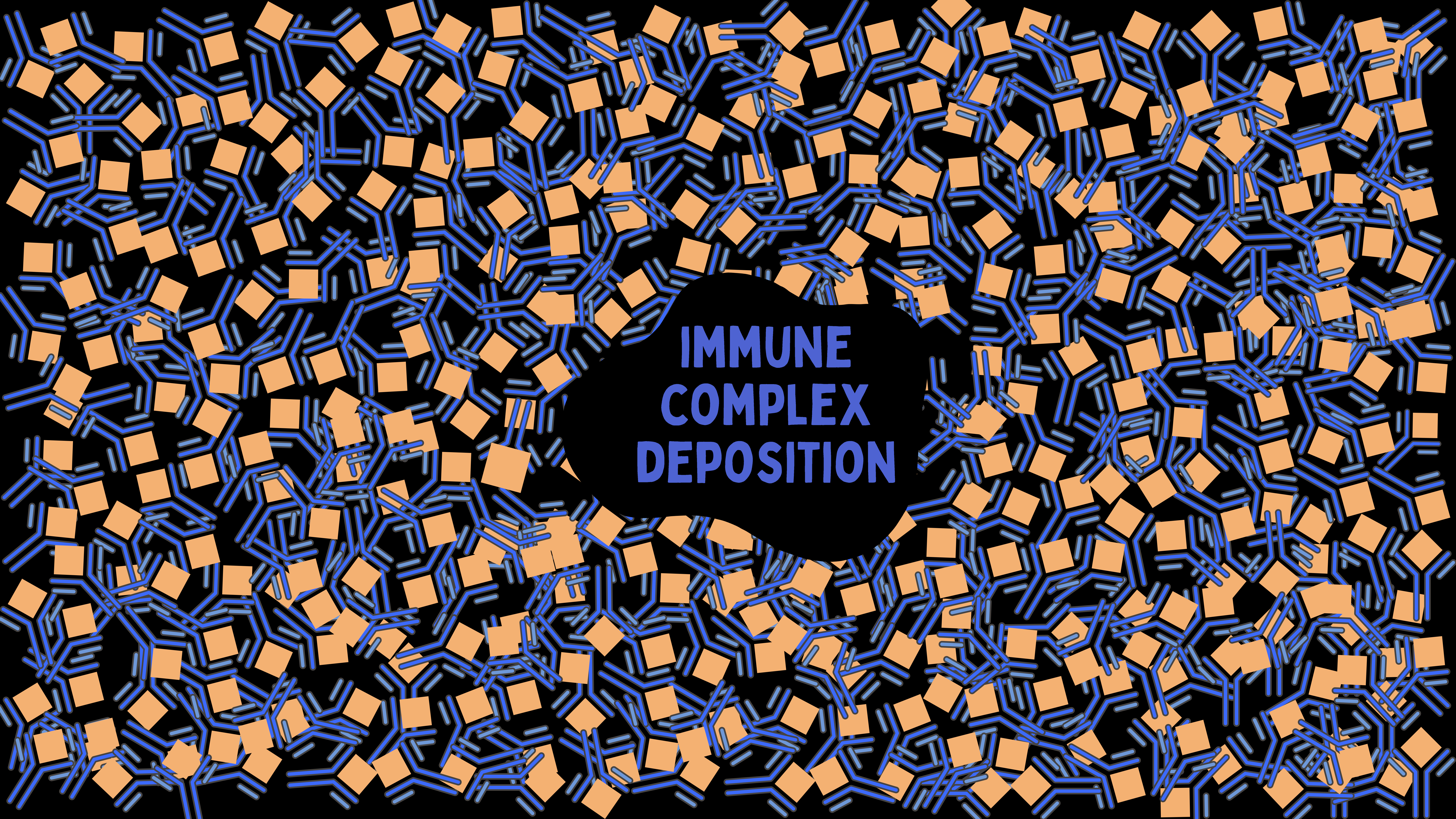
BECAUSE OF THE MEMORY CELLS...

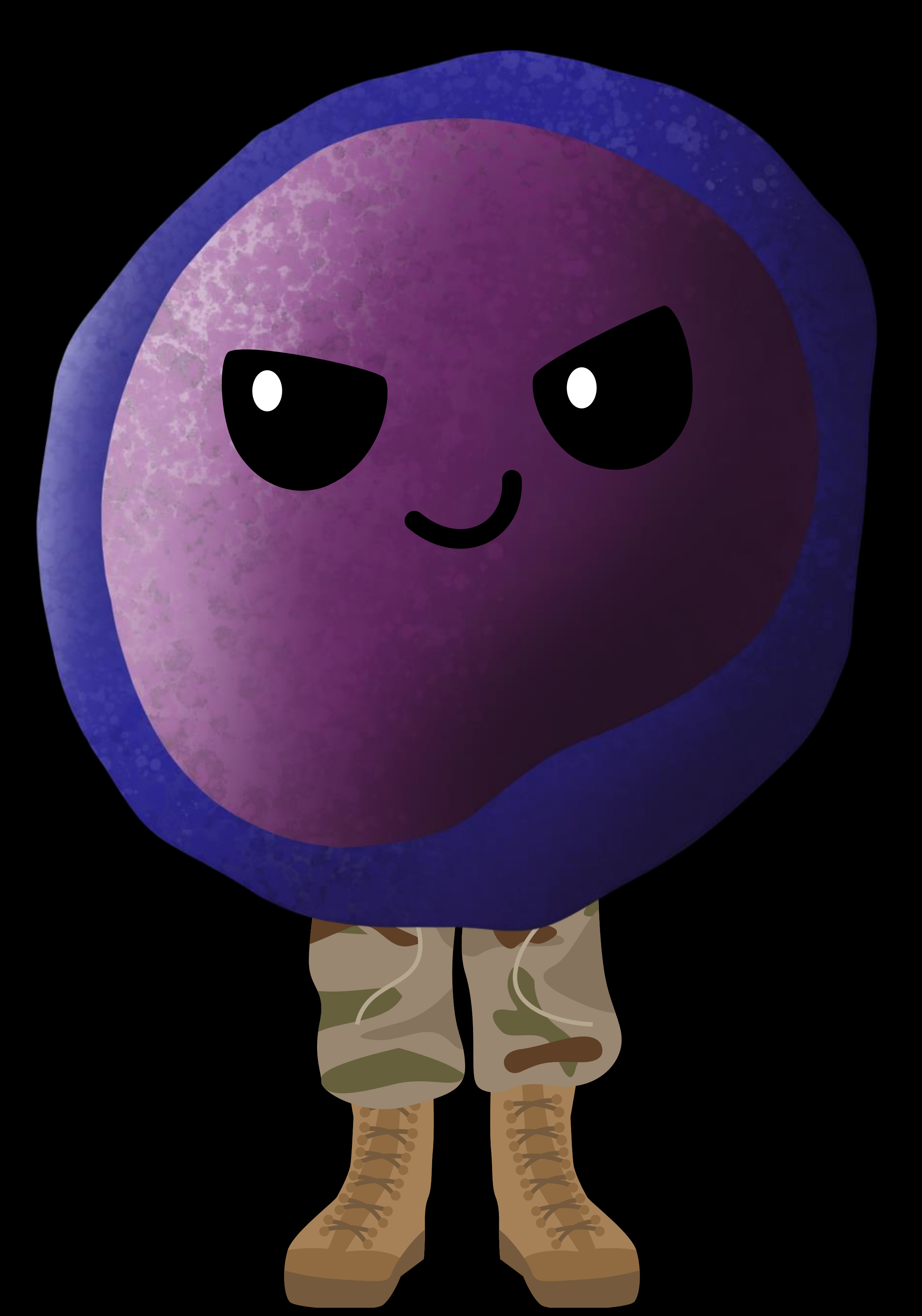
CONTINUED ATTACKS ON HEALTHY CELLS LEAD TO CELL DEATH AND ORGAN DAMAGE

COMPLEMENT

CYTOKINES

AUTOANTIBODIES


IMMUNE
COMPLEX
DEPOSITION



LUPUS

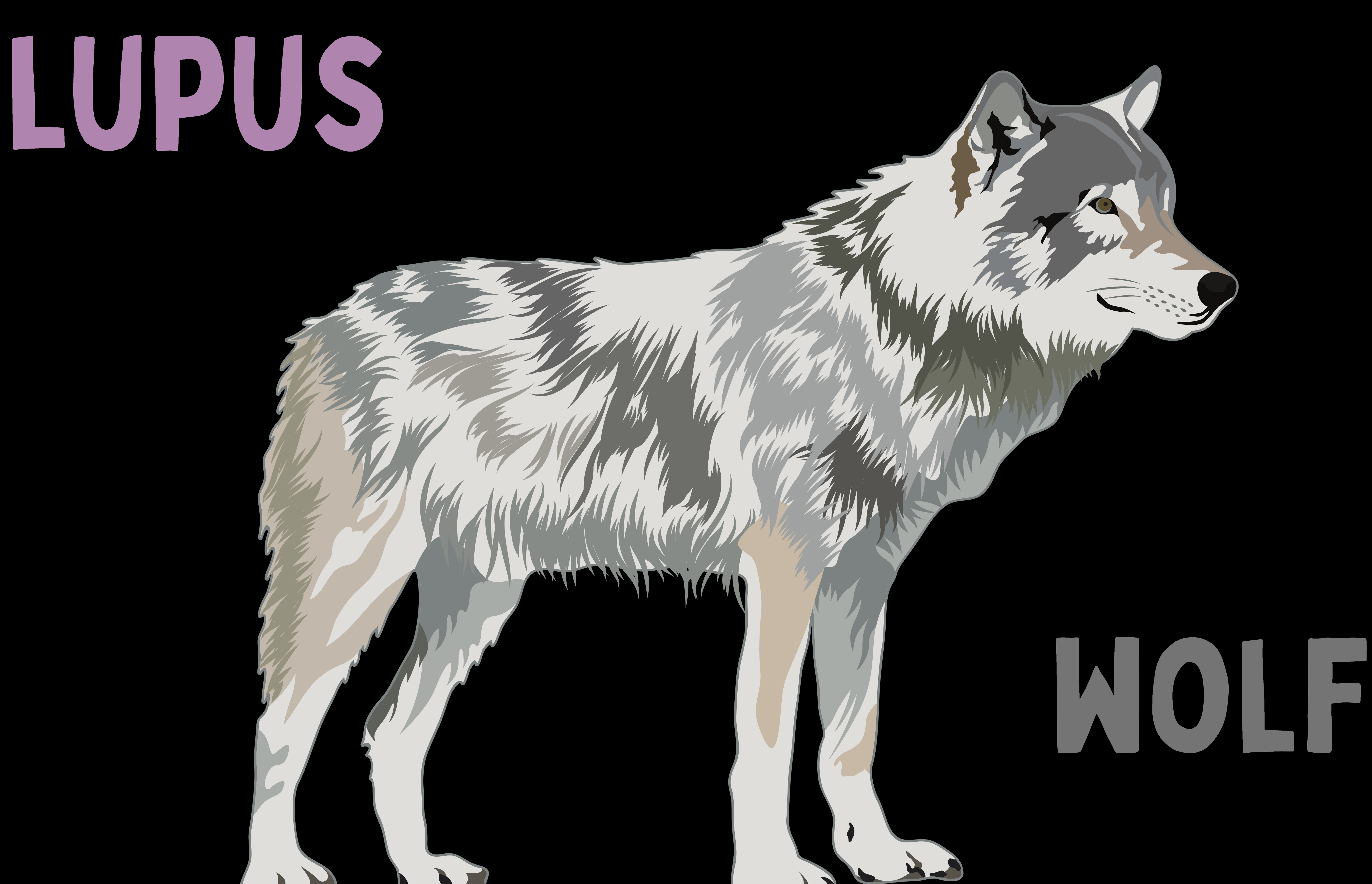
SCLERODERMA

REACTIVE ARTHRITIS

SYSTEMIC LUPUS ERYTHEMATOSIS

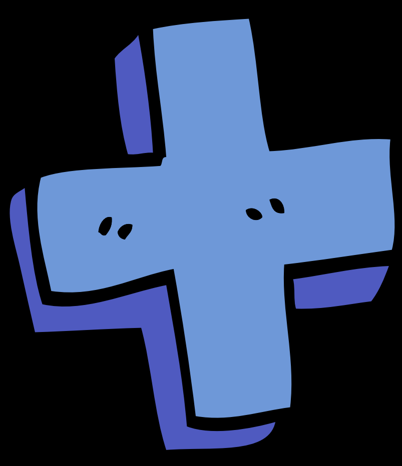
DRUG-INDUCED LUPUS
CUTANEOUS LUPUS
DISCOID LUPUS

LUPUS


LUPUS
WOLF

PROFESSOR LUPIN

GENETIC PREDISPOSITION



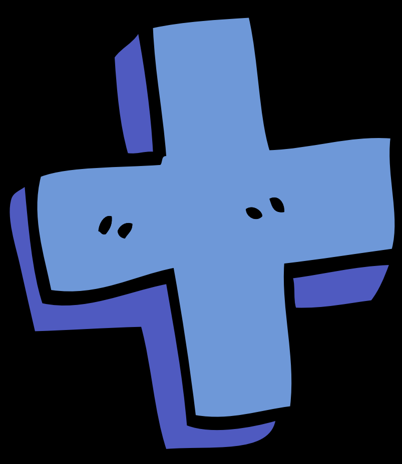
TRIGGER

INFECTION OR OTHER
ENVIRONMENTAL TRIGGER

WOMEN (46, XX) 10 TIMES RISK

KLINEFELTER SYNDROME (47, XXY) 14 TIMES RISK

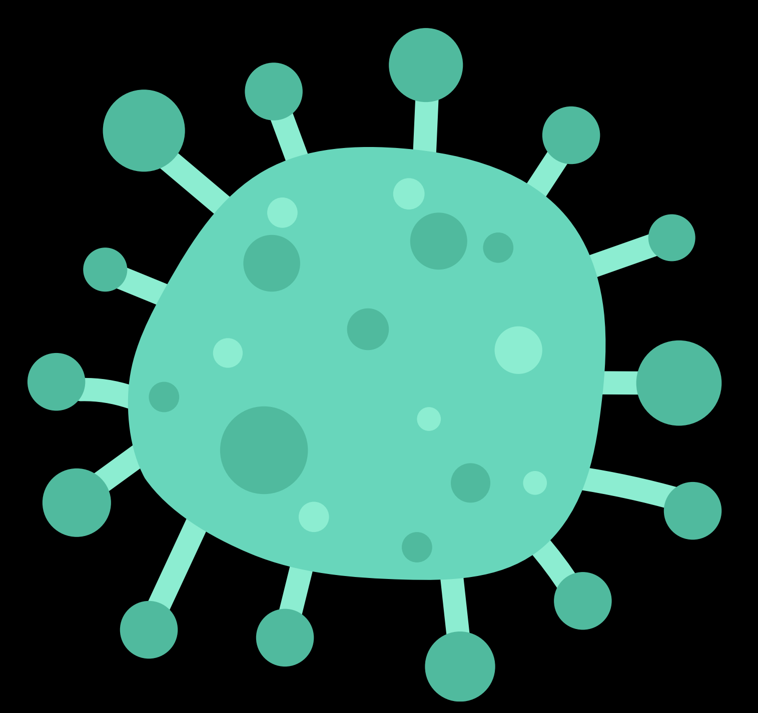
GENETIC PREDISPOSITION



TRIGGER

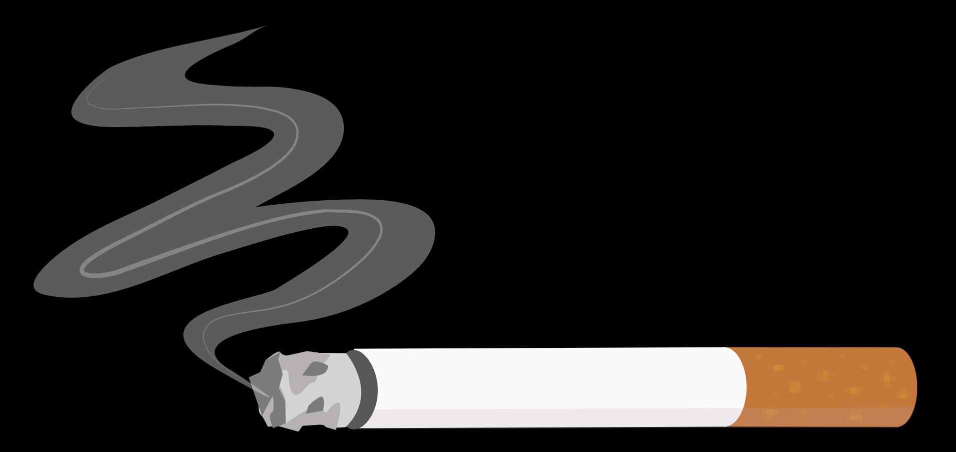
INFECTION OR OTHER
ENVIRONMENTAL TRIGGER

CELL DAMAGE



VIRUSES

UV LIGHT



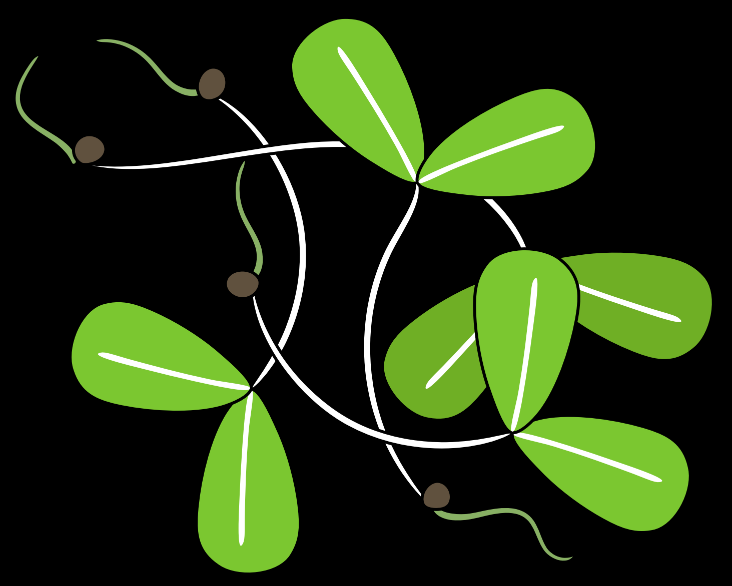
SMOKING

OTHER TRIGGERS

DRUGS

SULFA DRUGS
TETRACYCLINES
ESTROGEN

FOOD

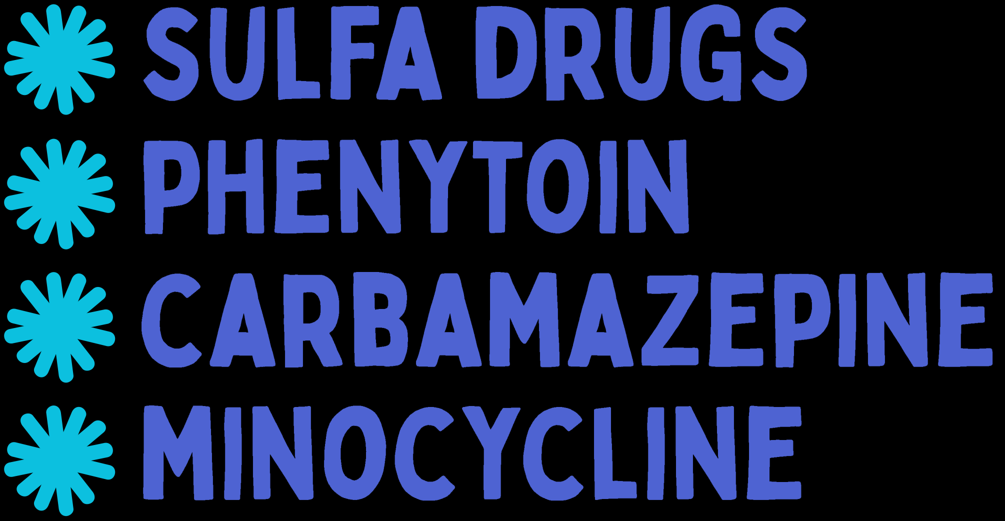


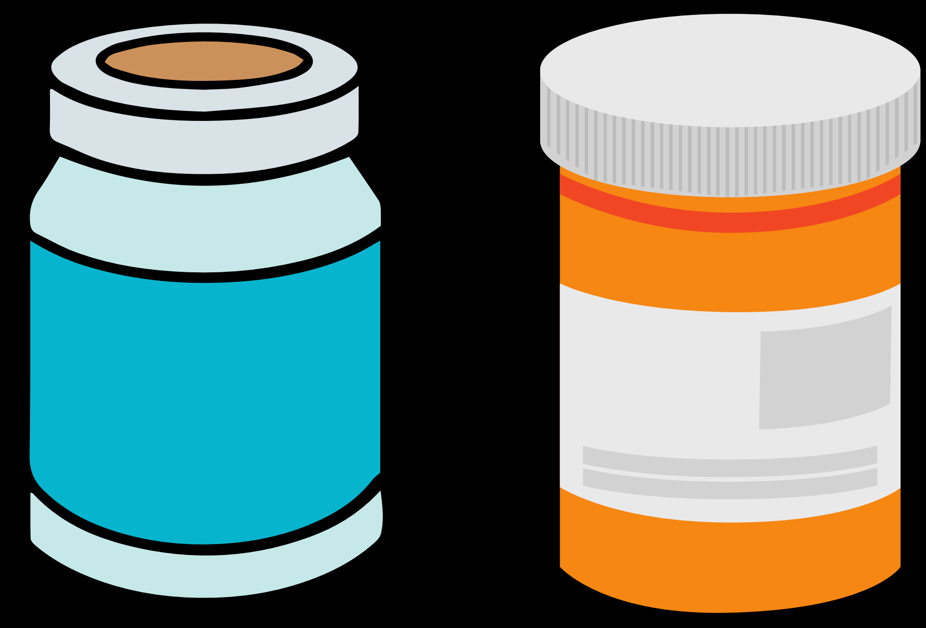
ALFALFA
SPROUTS

AMINO ACID
L-CANAVANINE

DRUG-INDUCED LUPUS ERYTHEMATOSIS

- LUPUS-LIKE SYNDROME DUE TO DRUG EXPOSURE
- IN A GENETICALLY SUSCEPTIBLE PERSON
- SYMPTOMS RESOLVE WHEN YOU STOP THE DRUG
- >100 DRUGS ON THE LIST

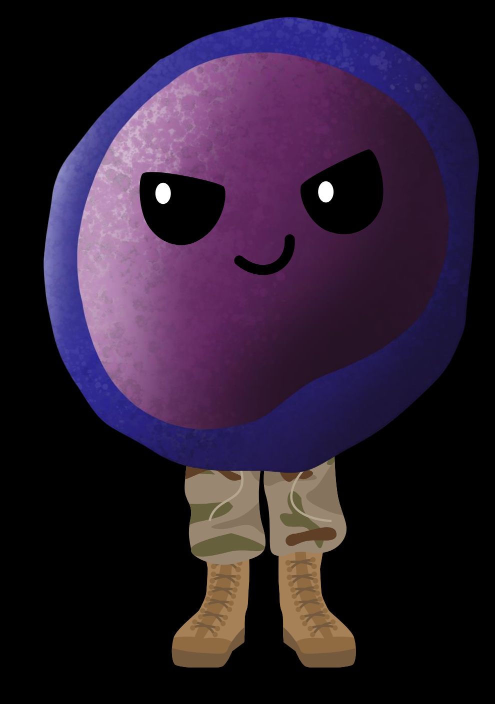




B CELLS AND T CELLS

PLASMA CELLS

MAKE ANTIBODIES THAT ATTACK HEALTHY CELLS



MEMORY B CELLS

REMEMBER PATHOGENS FOR NEXT TME

HELPER T CELLS

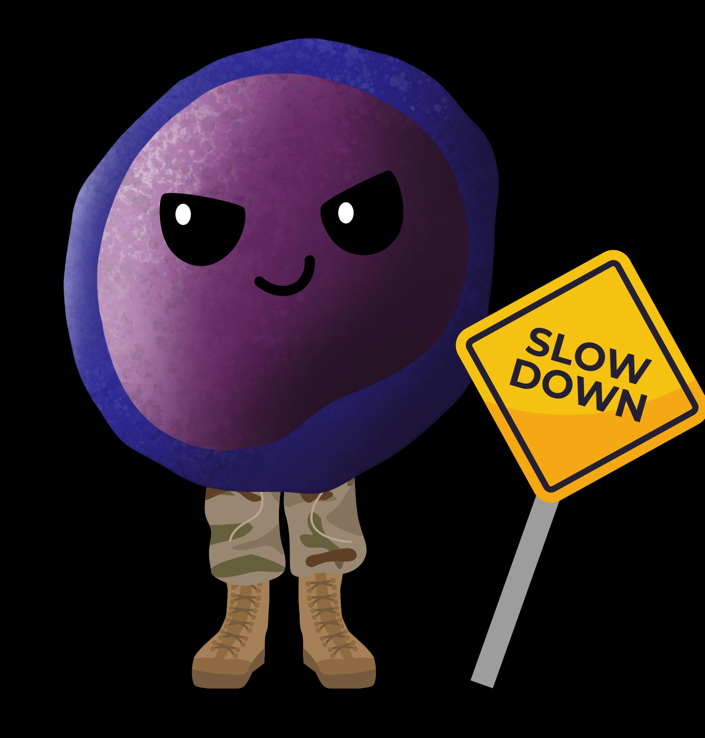
SOUND THE ALARM WITH CYTOKINES

KILLER T CELLS

KILL HEALTHY CELLS

MEMORY T CELLS

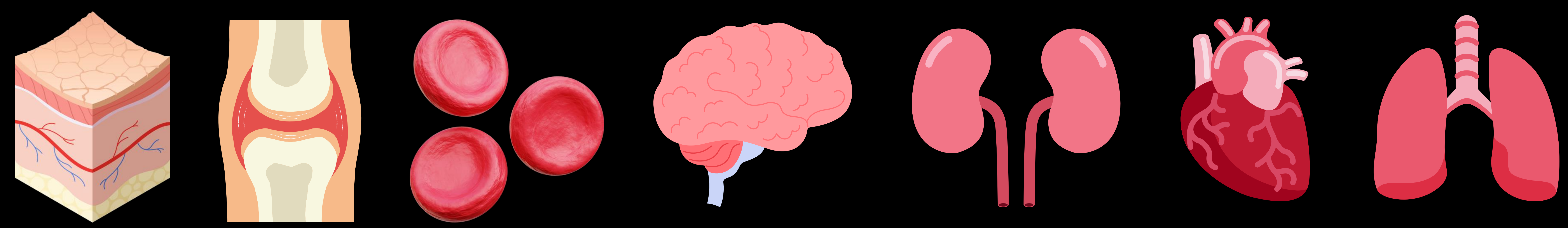
AUTOREACTIVE CELLS KEEP CONSTANT INFLAMMATORY RESPONSE



REGULATORY T CELLS

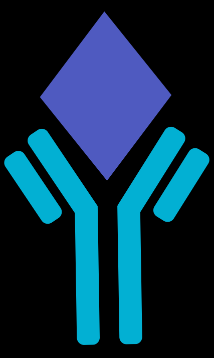
FEWER CELLS PRESENT

LUPUS CAN AFFECT ANY ORGAN SYSTEM

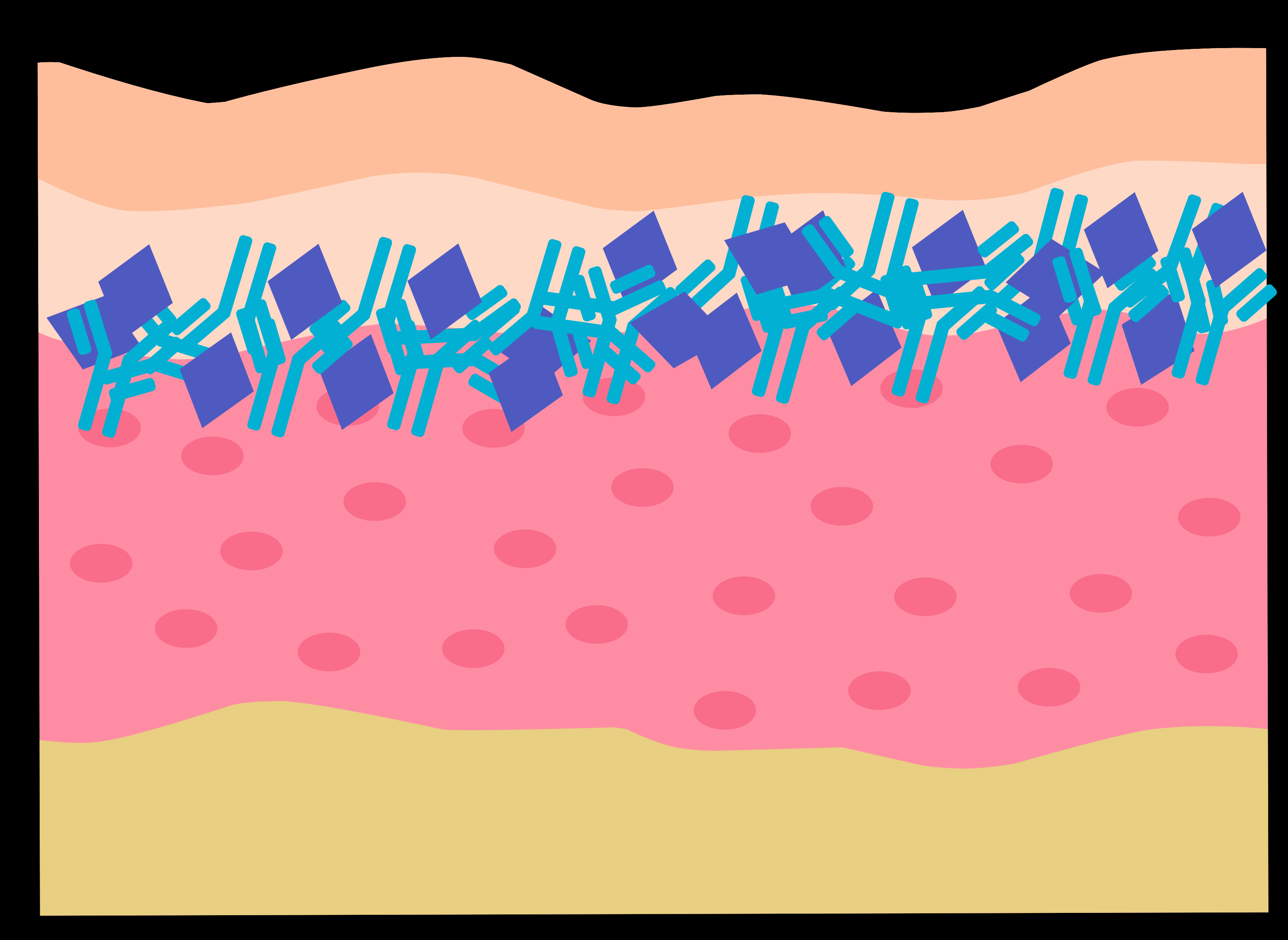


⭐ INCLUDE LUPUS FLARE IN YOUR DIFFERENTIAL FOR ALMOST ANY CHIEF COMPLAINT IN PATIENTS WITH SLE

80% OF PATIENTS WITH SLE WILL HAVE A SKIN PROBLEM



IMMUNE COMPLEX DEPOSITION AT THE DERMAL-EPIDERMAL JUNCTION



LUPUS ERYTHEMATOSIS

SYSTEMIC LUPUS ERYTHEMATOSIS (ACUTE CUTANEOUS LE)

- GENERALIZED ACLE
- LOCALIZED ACLE

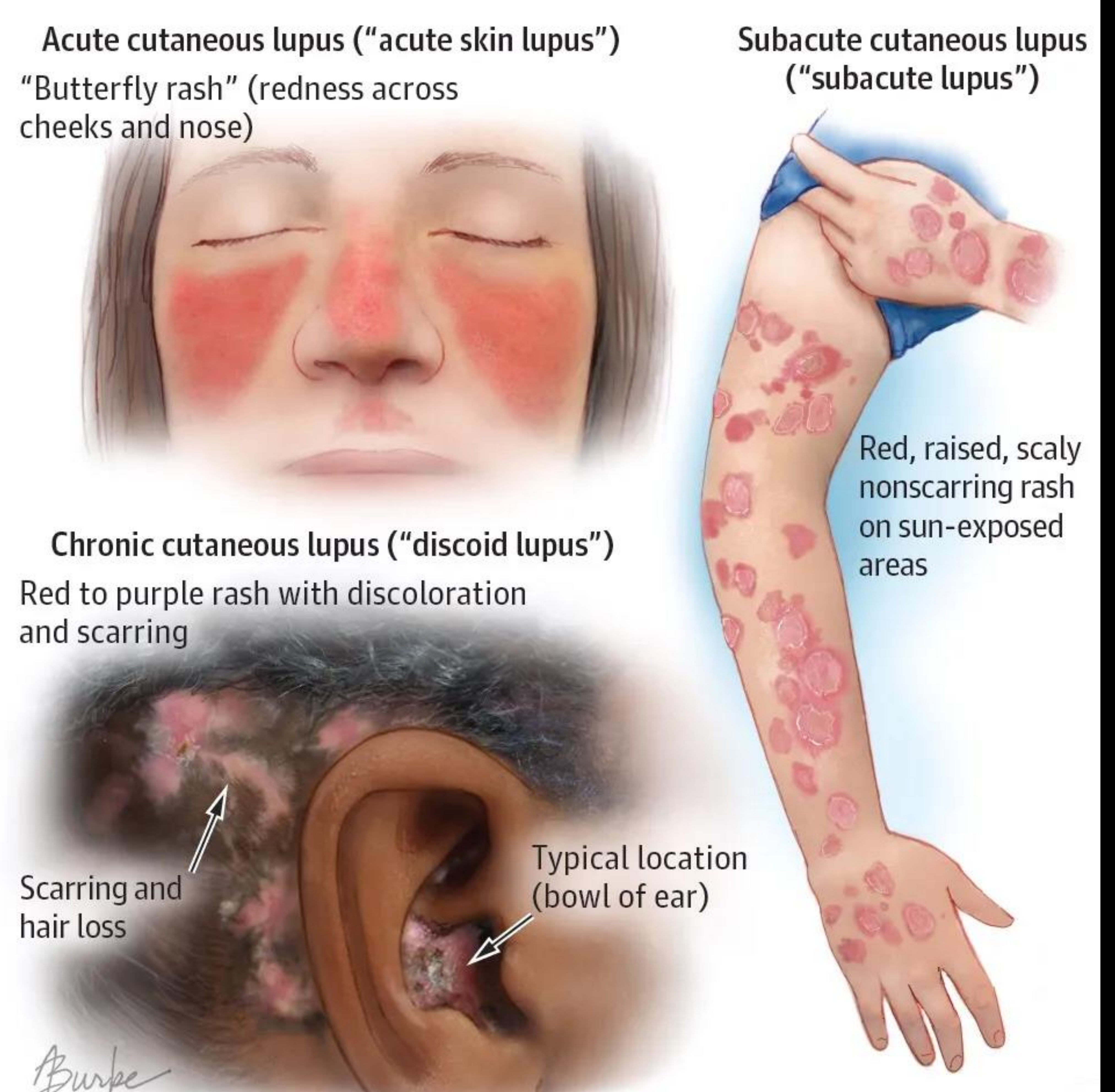
SUBACUTE CUTANEOUS LE (SCLE)

- PAPULOSQUAMOUS SCLE
- ANNULAR SCLE

CHRONIC CUTANEOUS LE

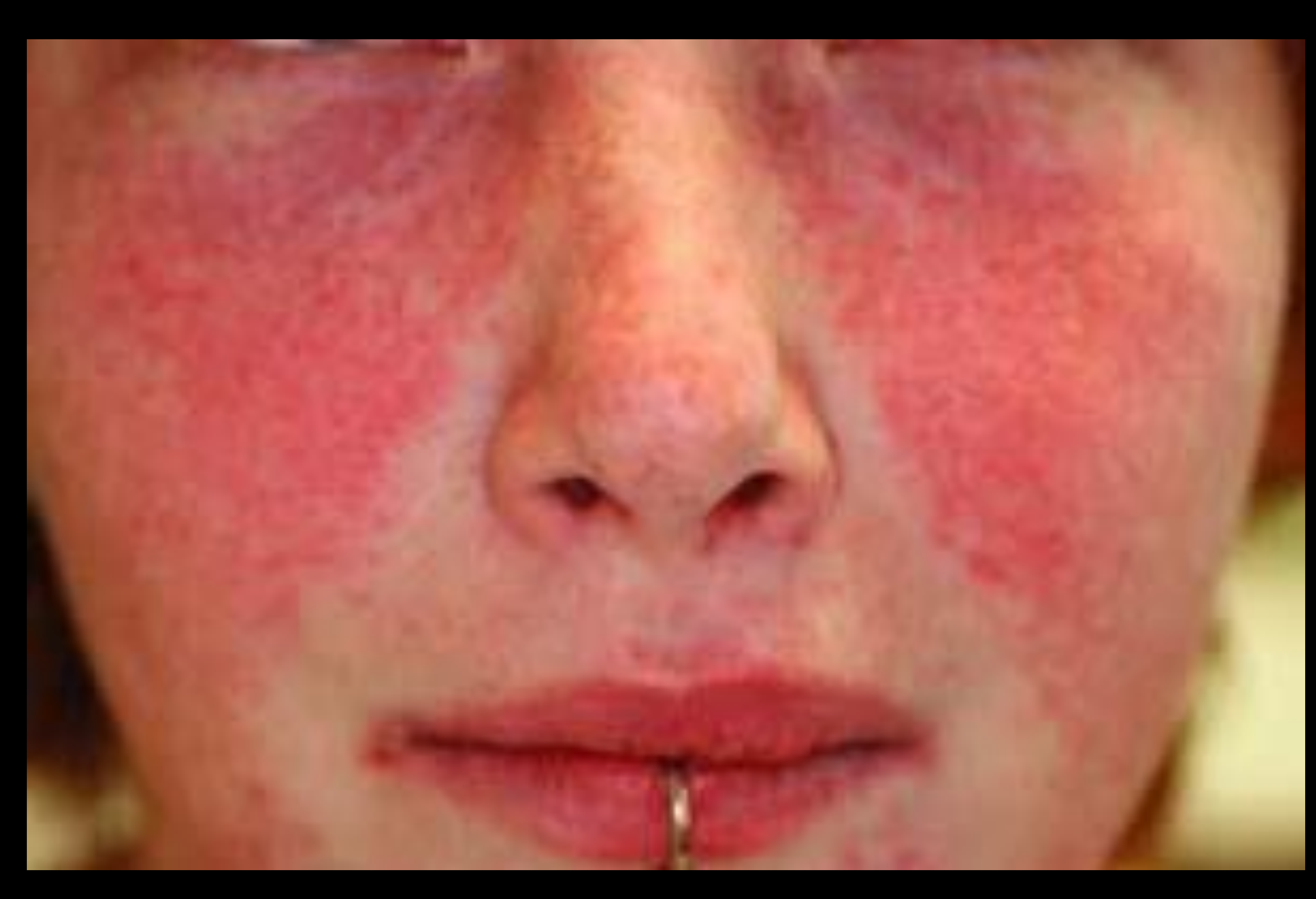
- DISCOID LUPUS (DLE)
- LICHENOID DLE
- MUCOSAL DLE
- HYPERTROPHIC
- CHILBLAIN LE
- LUPUS PROFUNDUS
- LUPUS TIMIDUS

SUN-EXPOSED AREAS OF SKIN

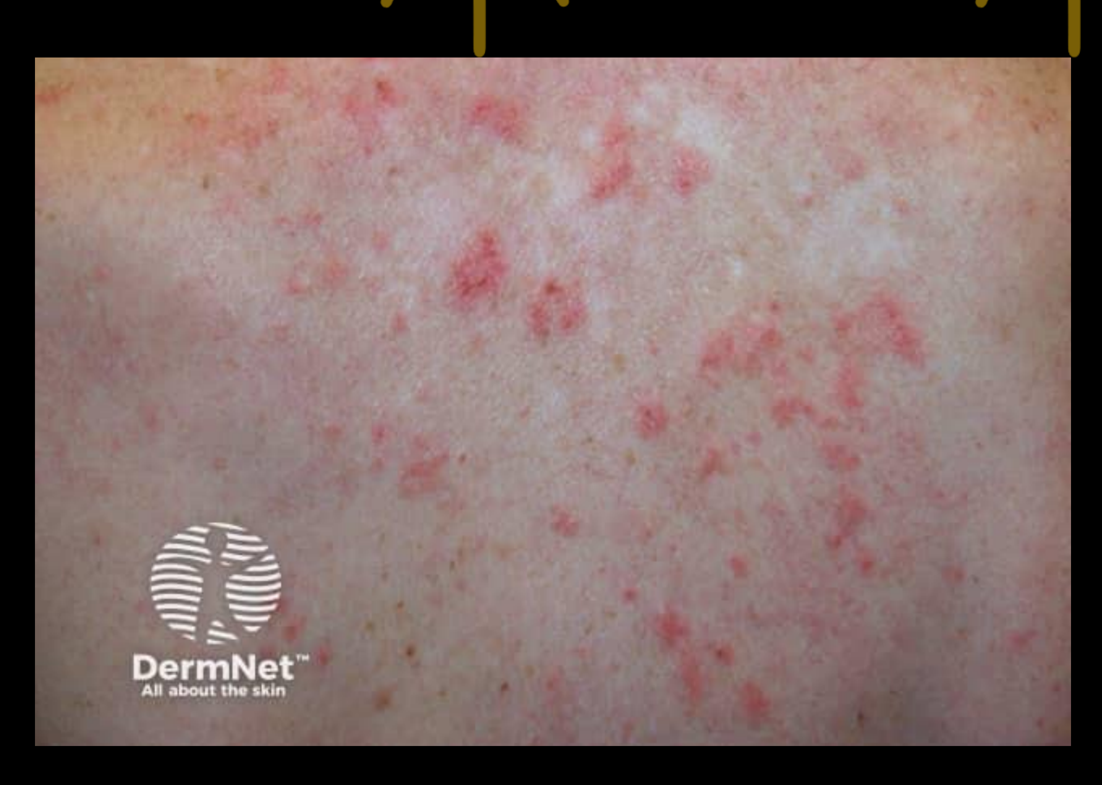


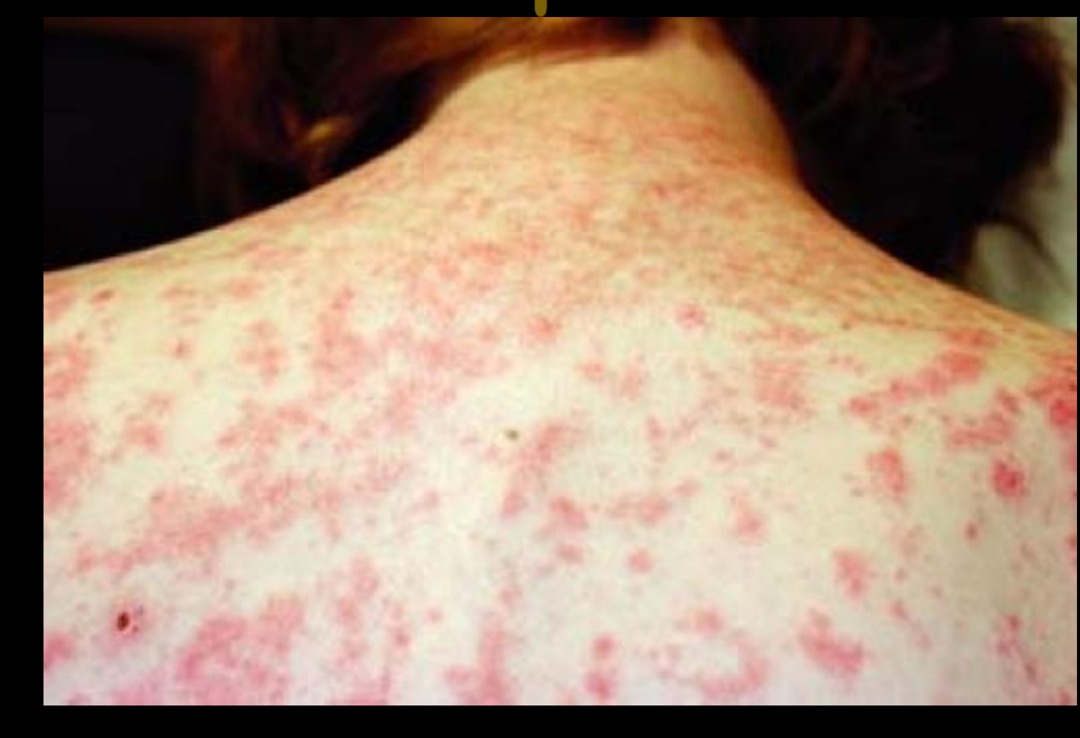
Types of cutaneous lupus erythematosus
Acute cutaneous lupus ("acute skin lupus")
"Butterfly rash" (redness across cheeks and nose)
Subacute cutaneous lupus ("subacute lupus")
Red, raised, scaly nonscarring rash on sun-exposed areas
Chronic cutaneous lupus ("discoid lupus")
Red to purple rash with discoloration and scarring
Scarring and hair loss
Typical location (bowl of ear)
Burke

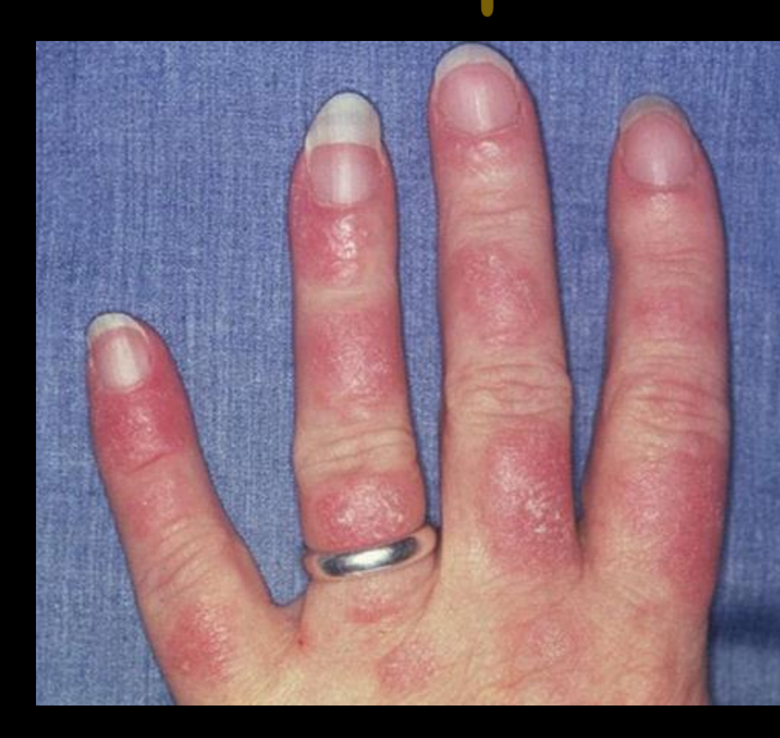
THE MALAR RASH OF LUPUS SPARES THE NASOLABIAL FOLDS

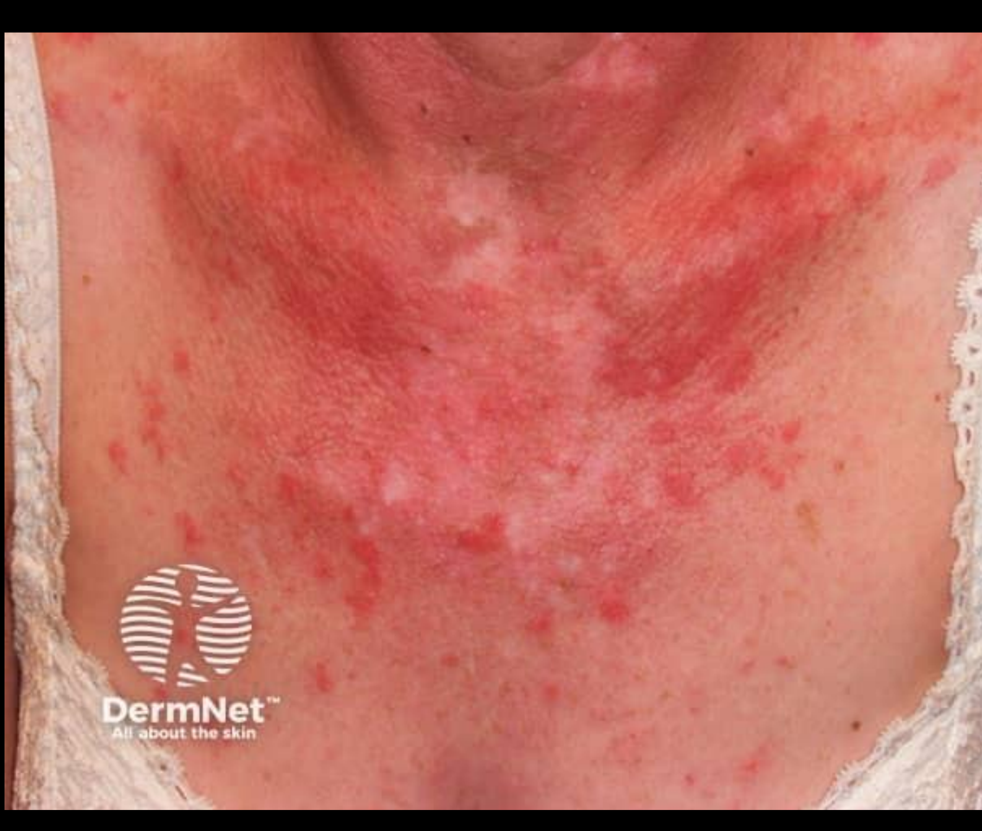


SUBACUTE PHOTOSENSITIVE LUPUS RASHES

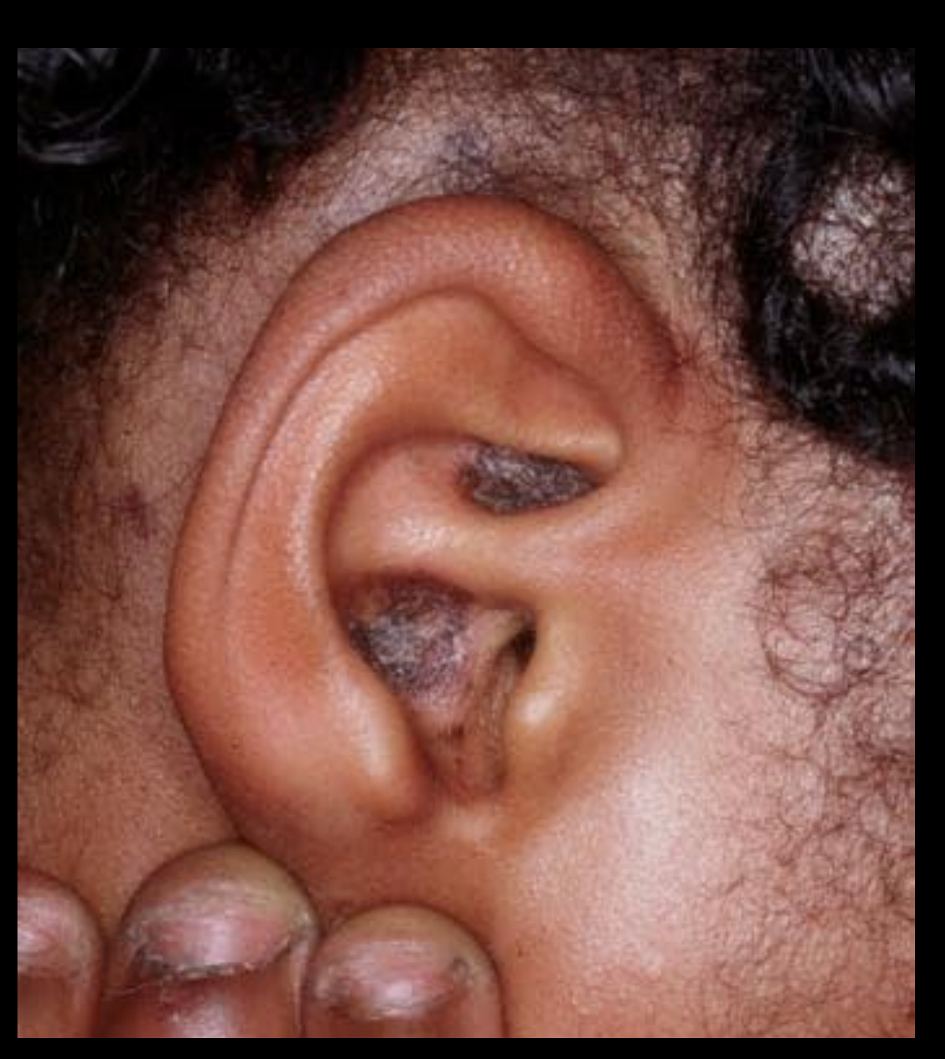
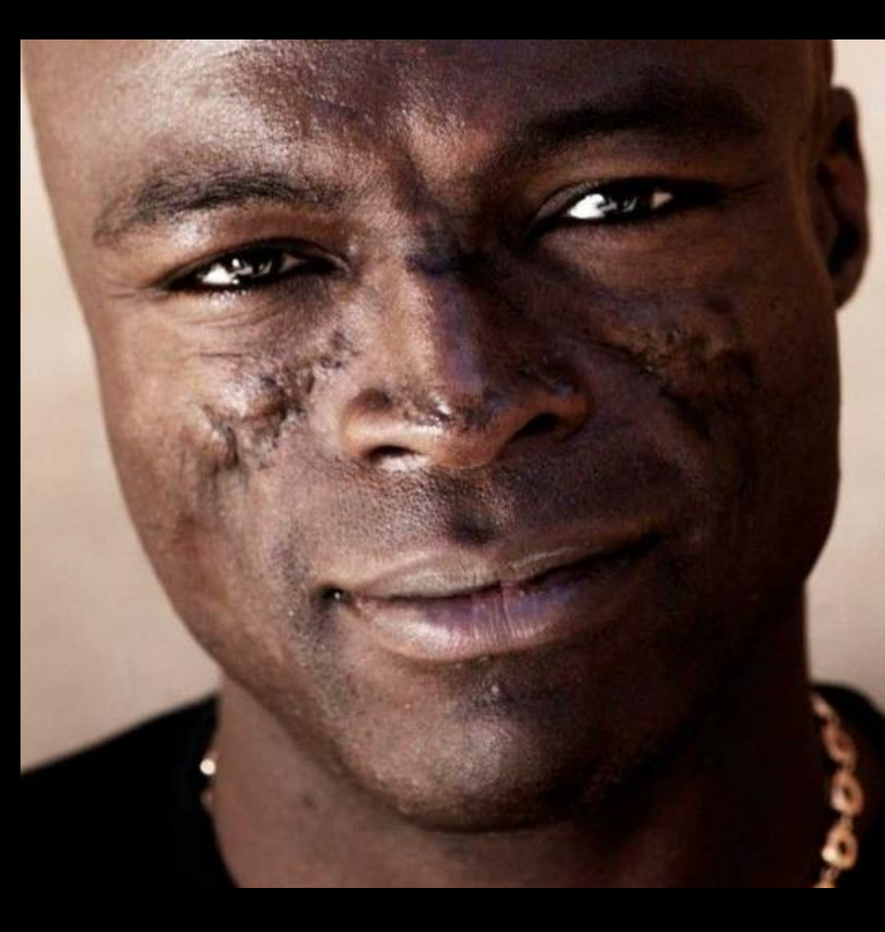
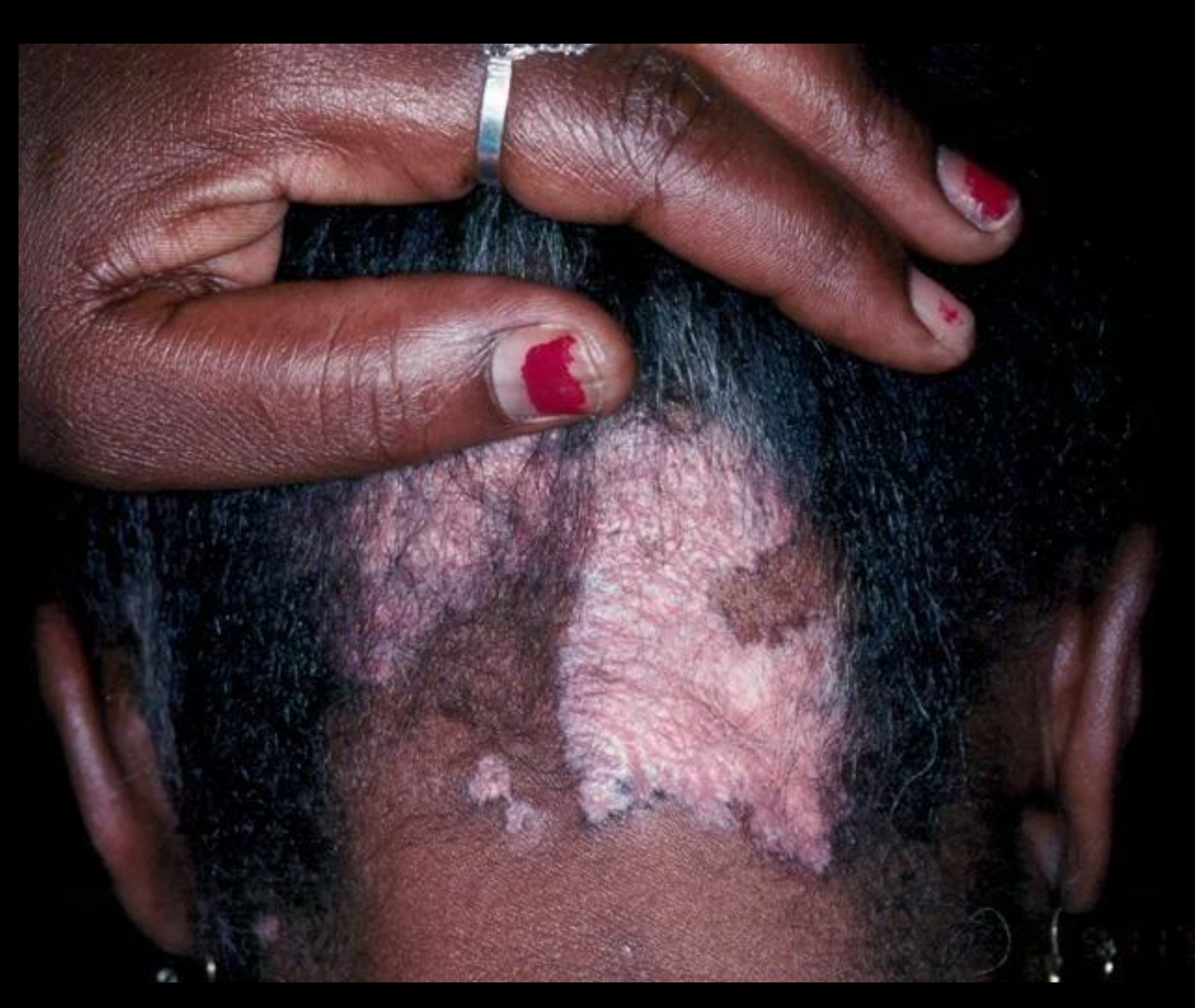
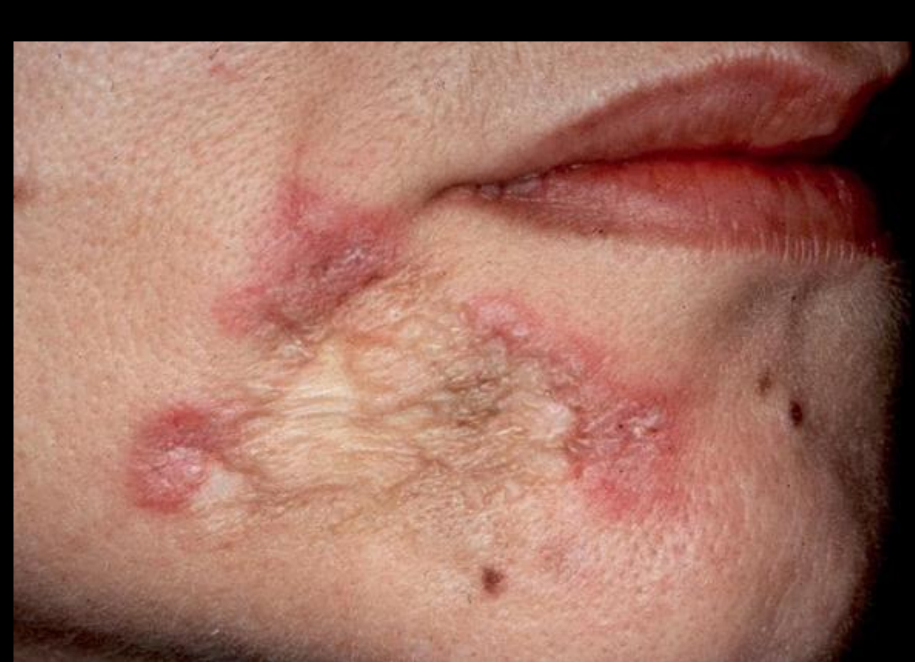
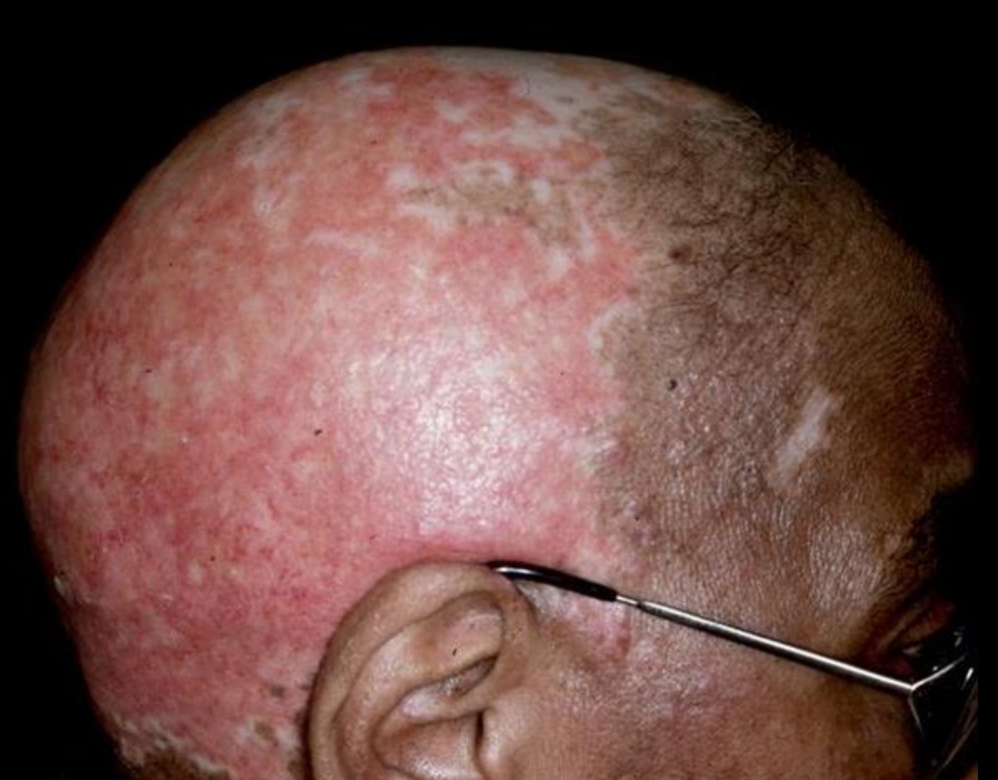
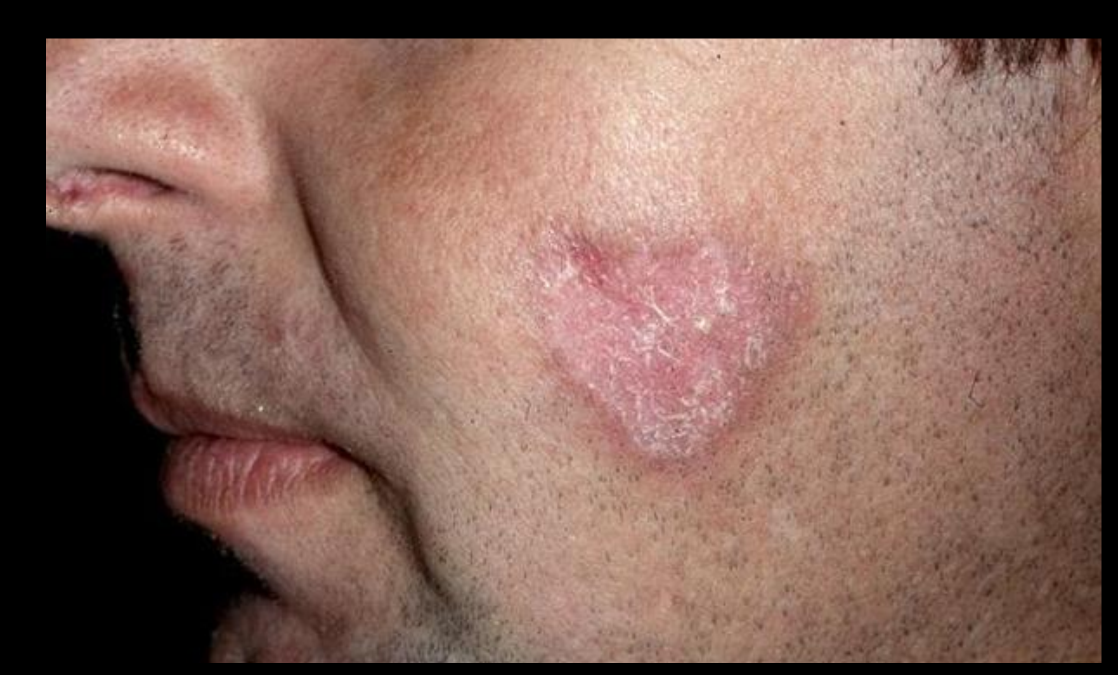


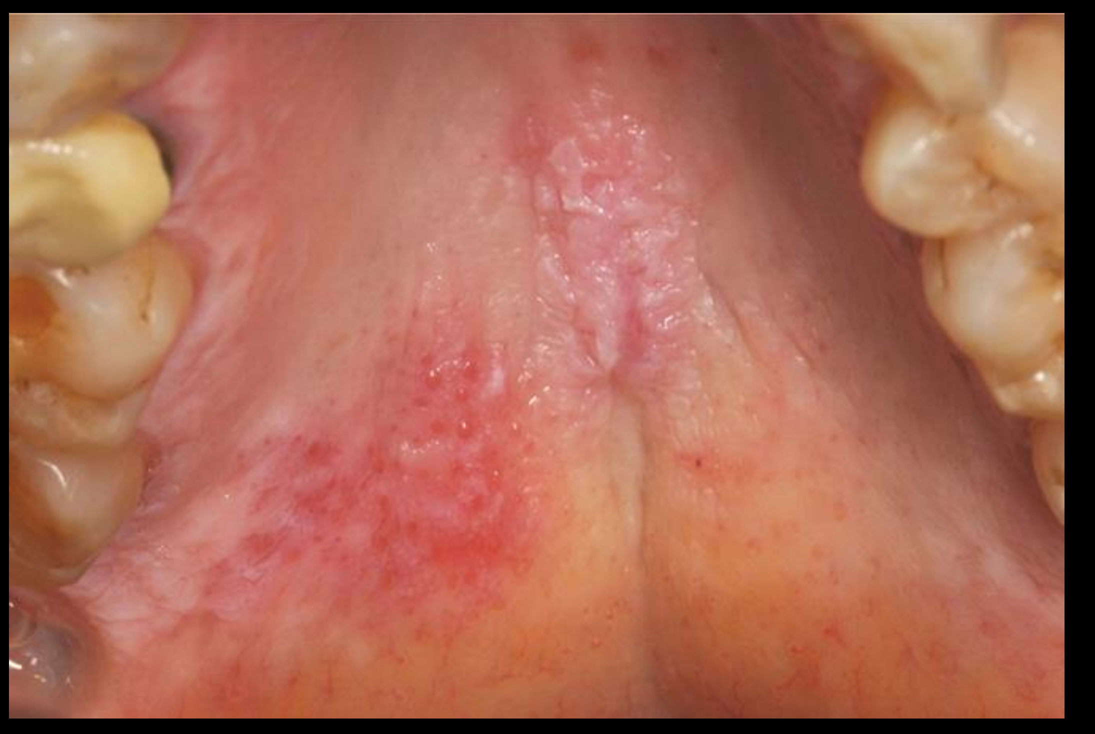


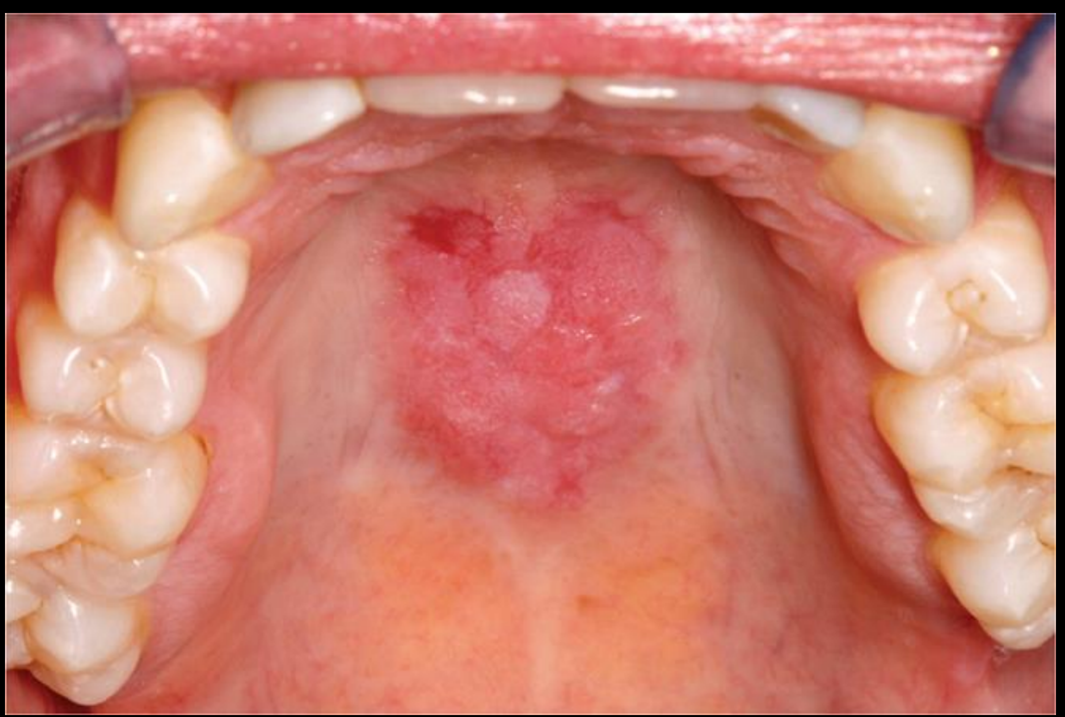


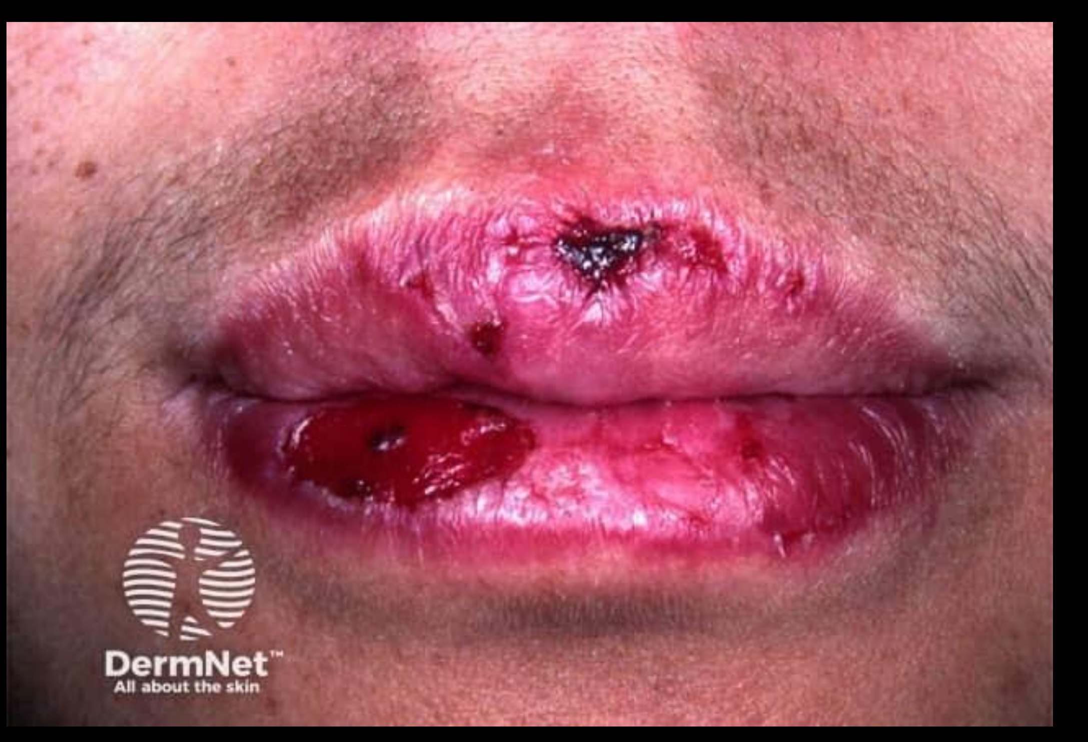


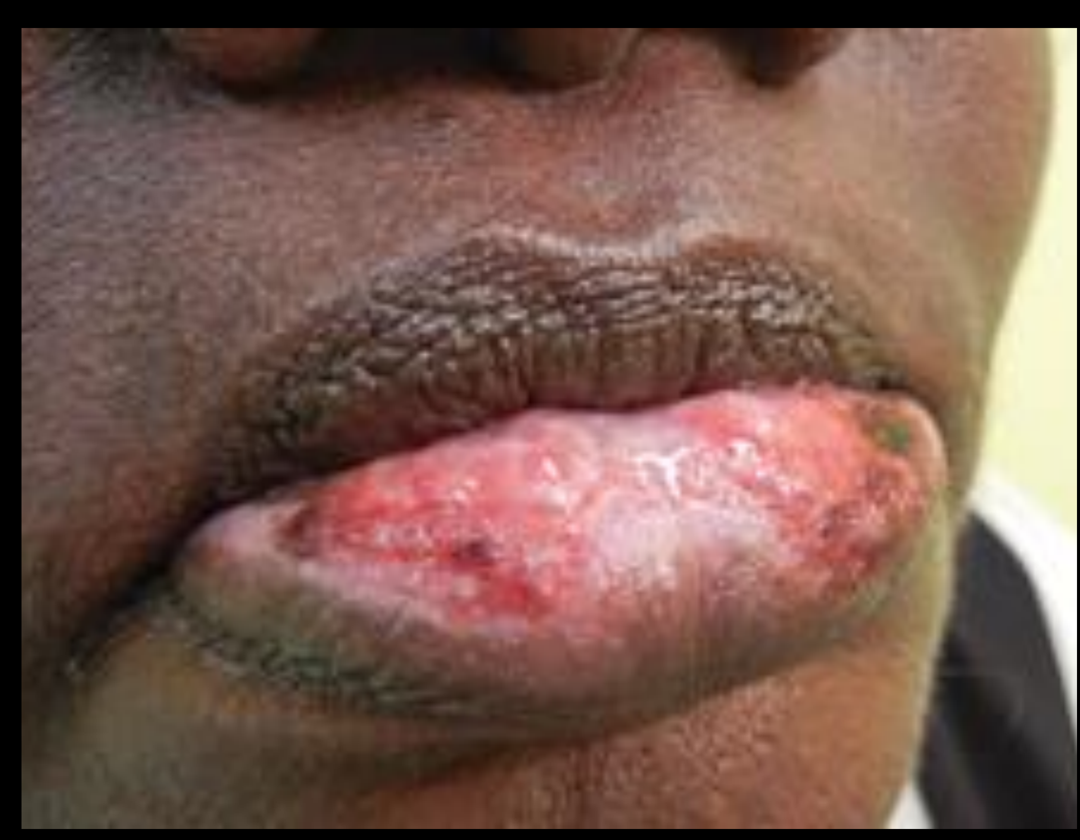
CHRONIC CUTANEOUS DISCOID LUPUS

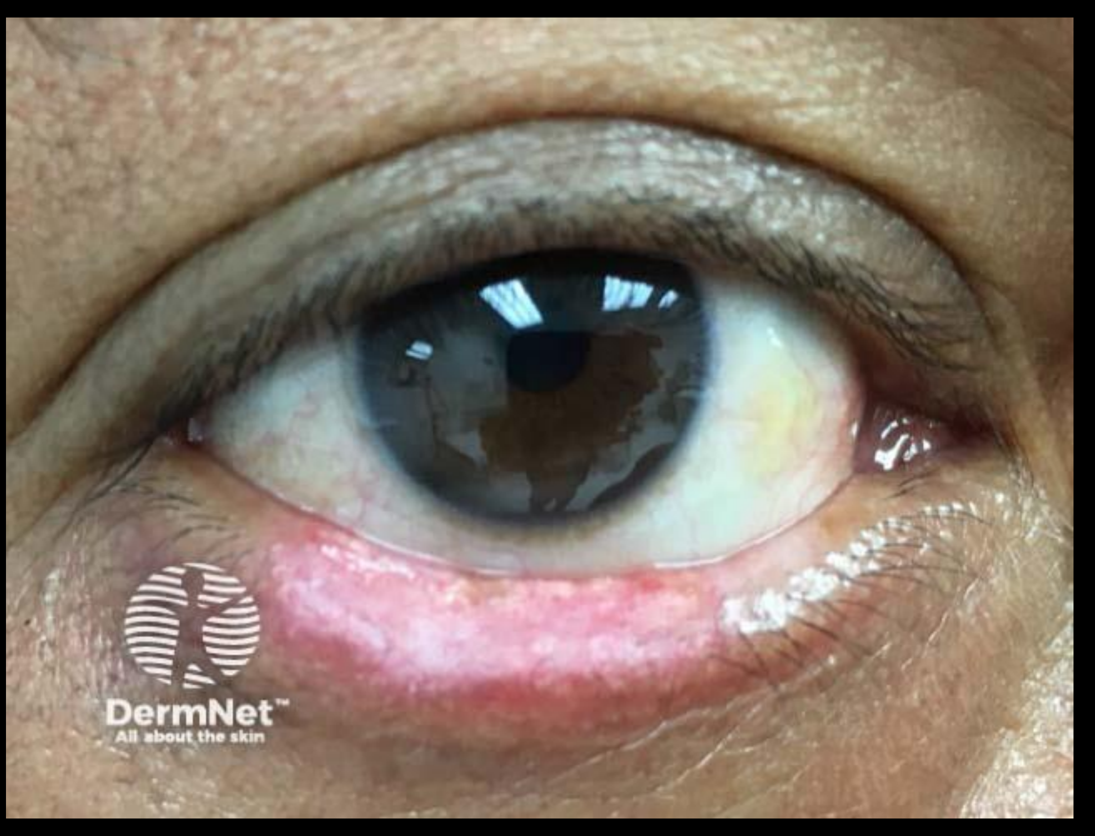












DermNet™
All about the skin

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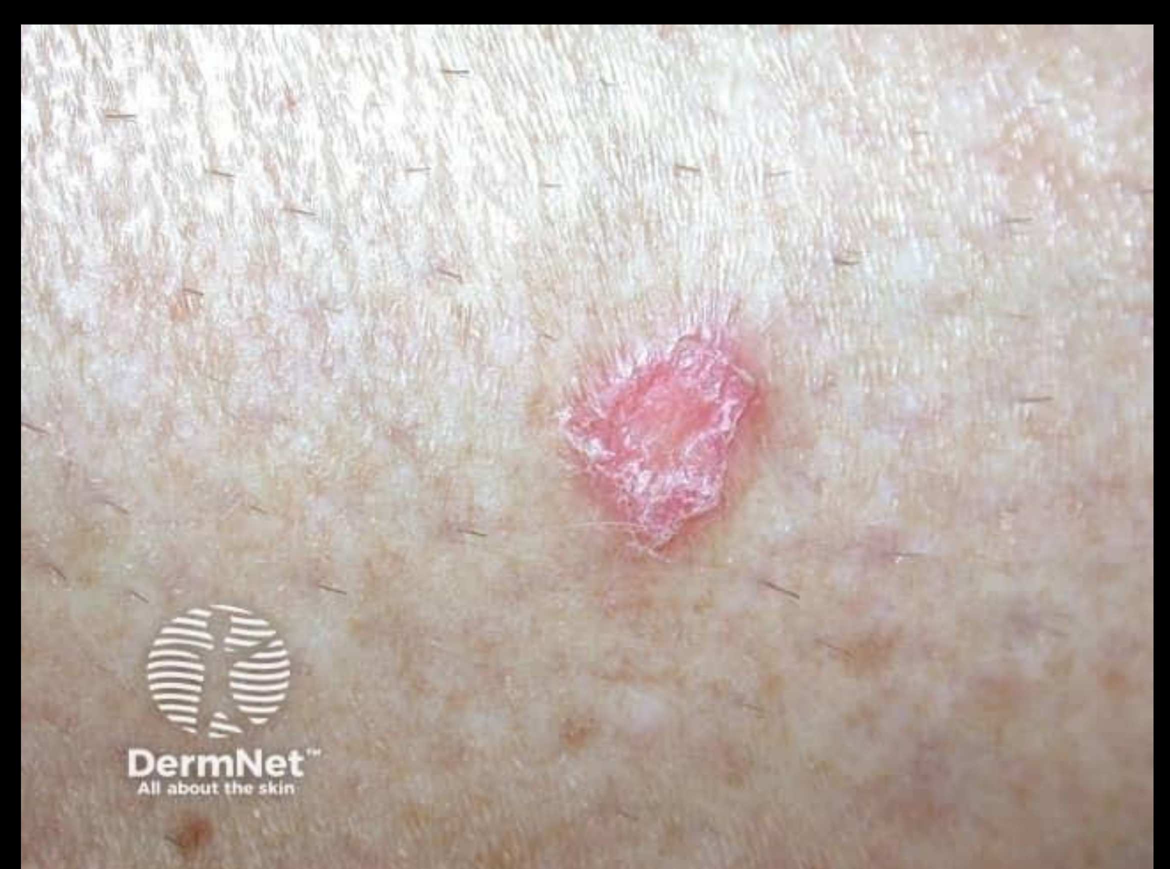
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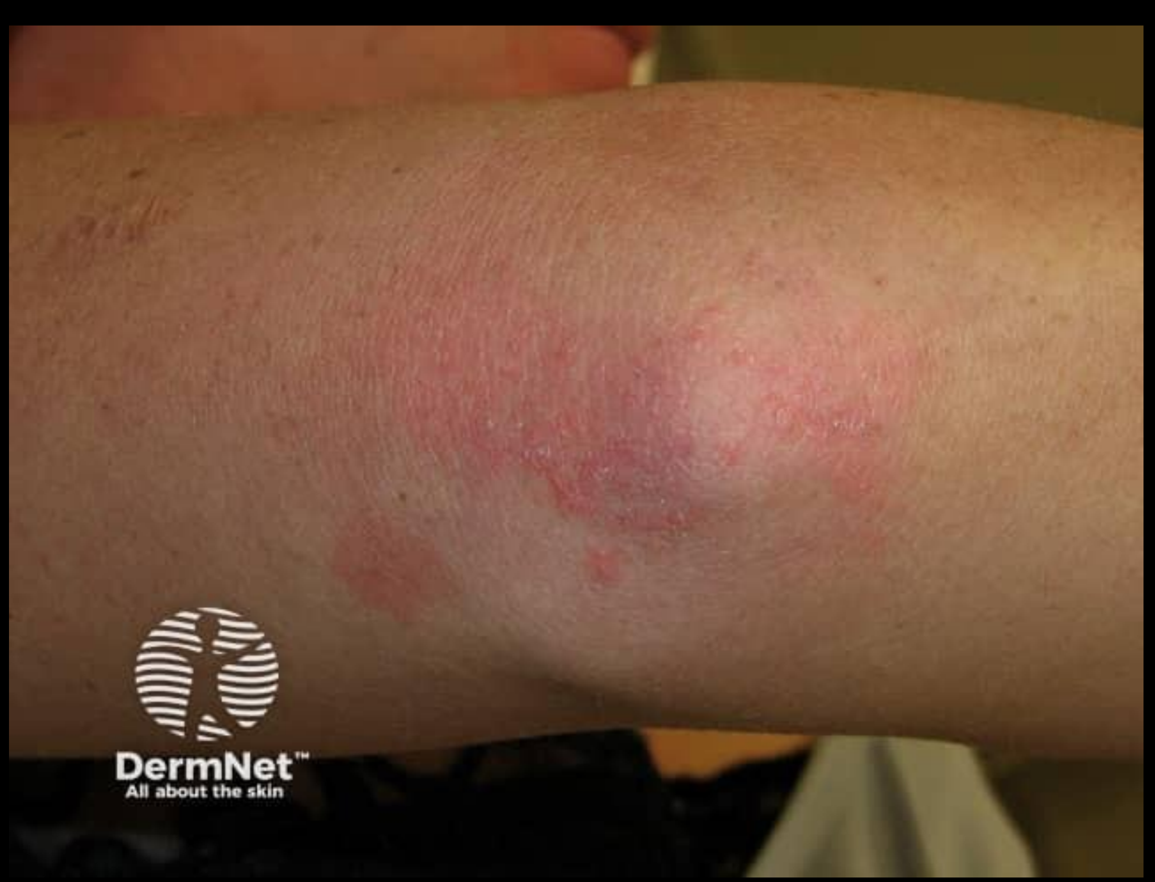
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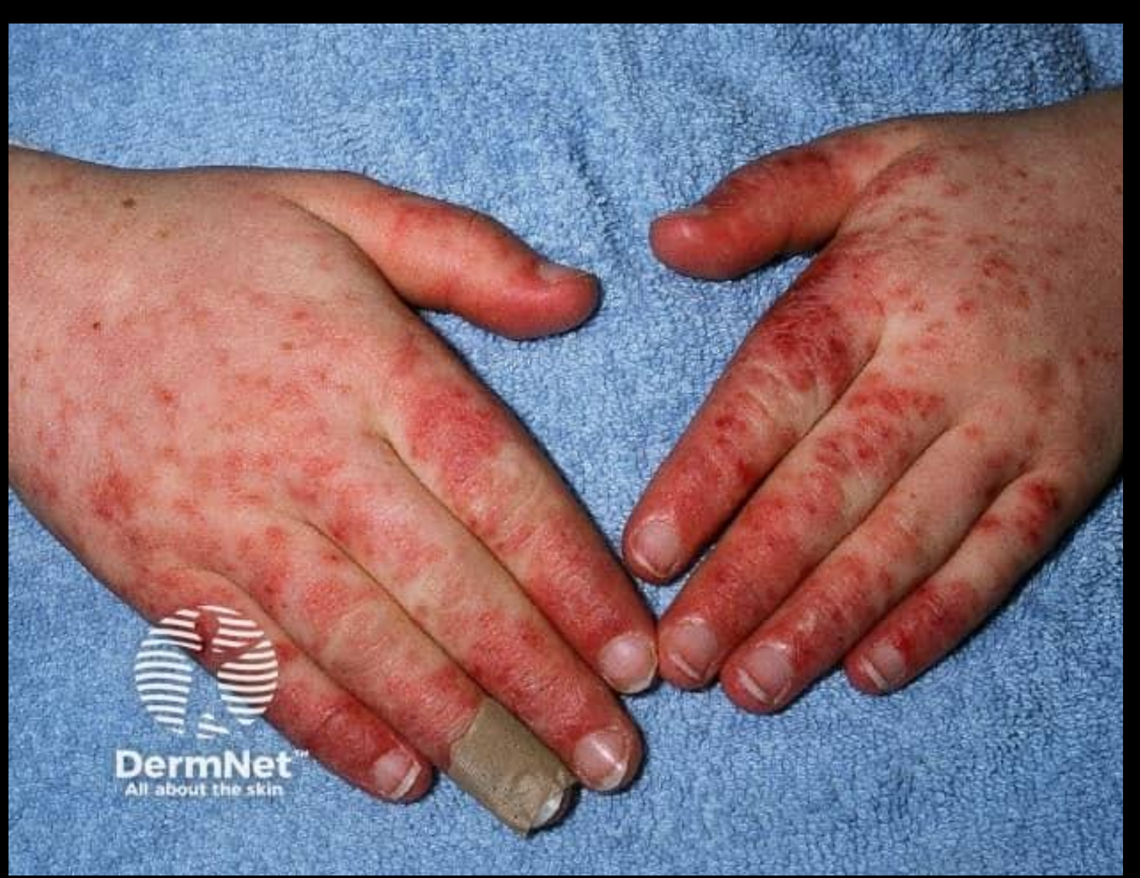
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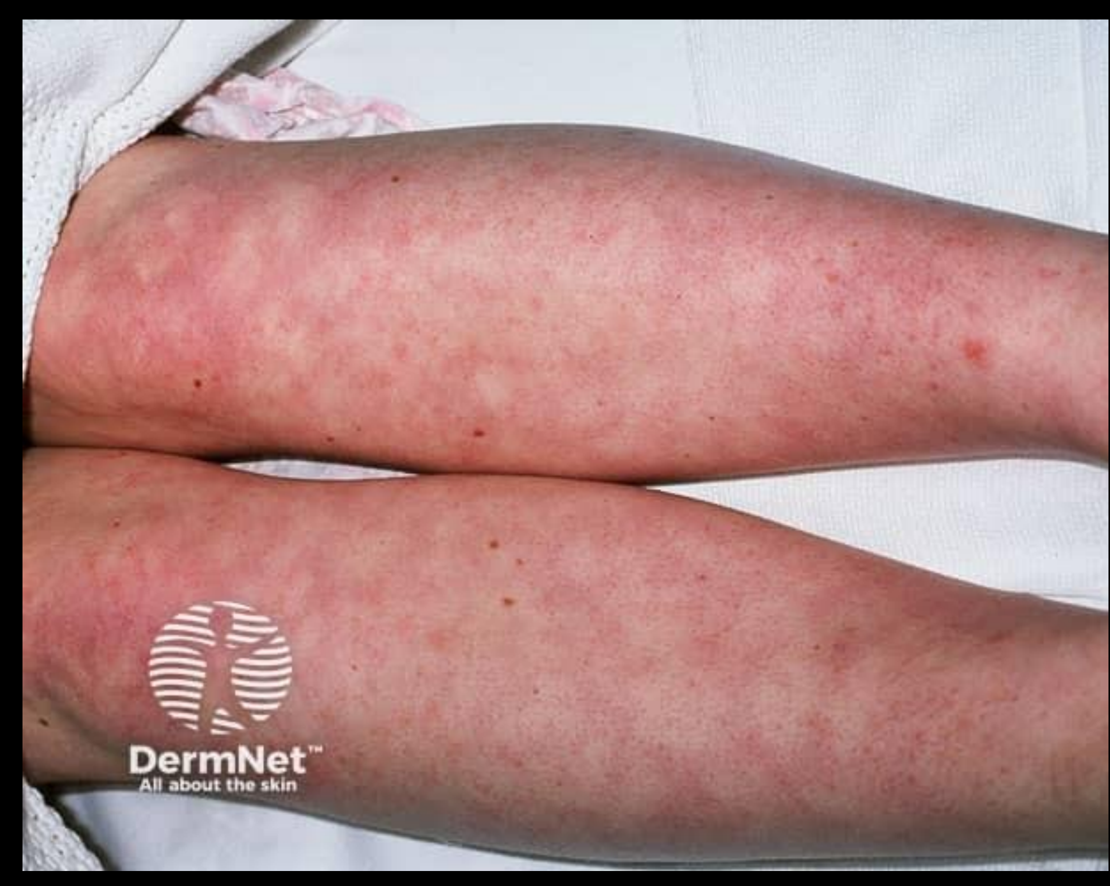
DermNet™
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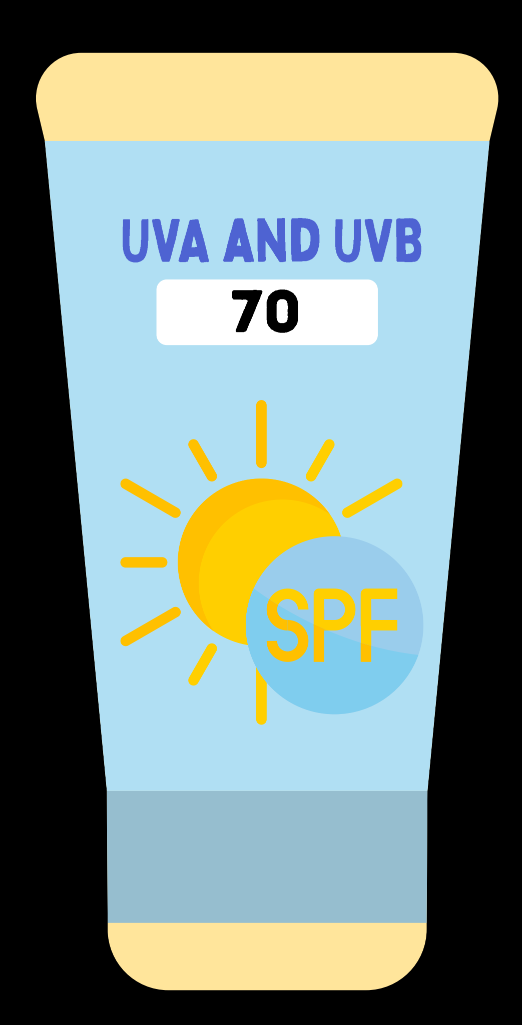
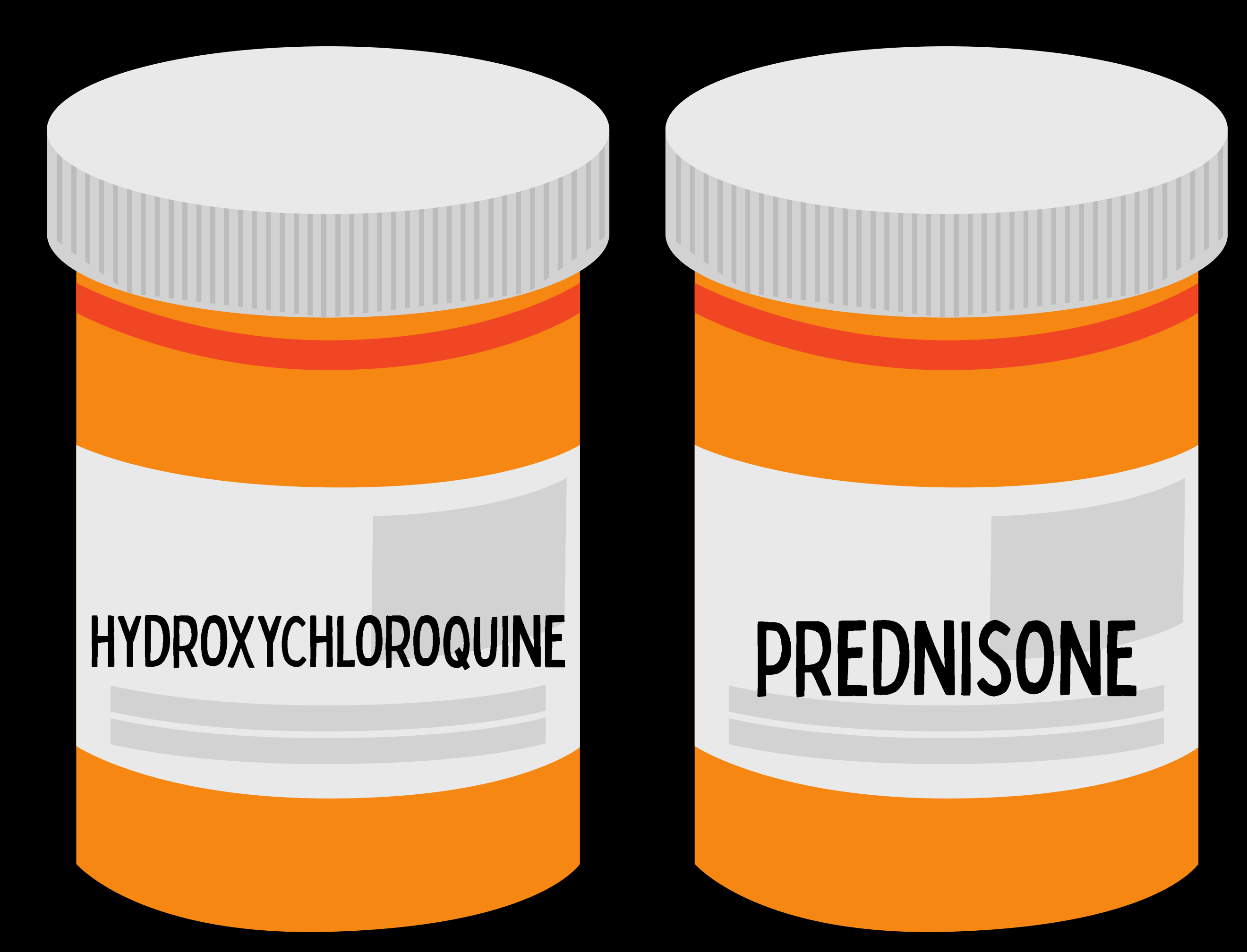
TREATMENT OF CUTANEOUS LUPUS



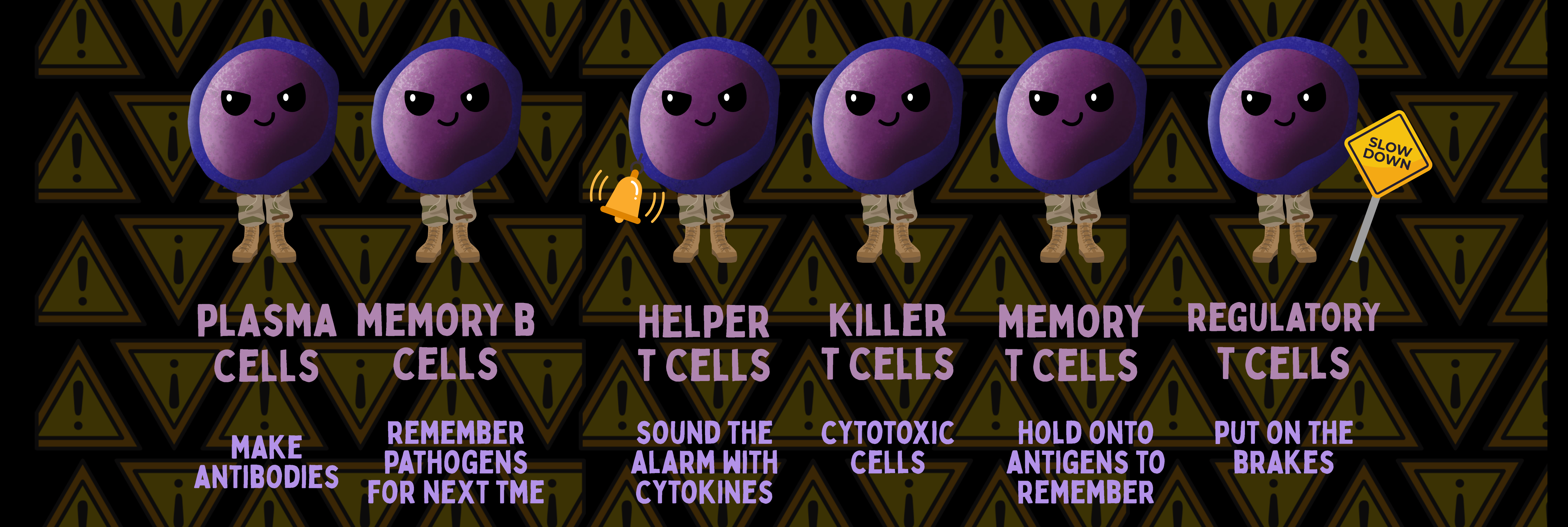
SUNSCREEN



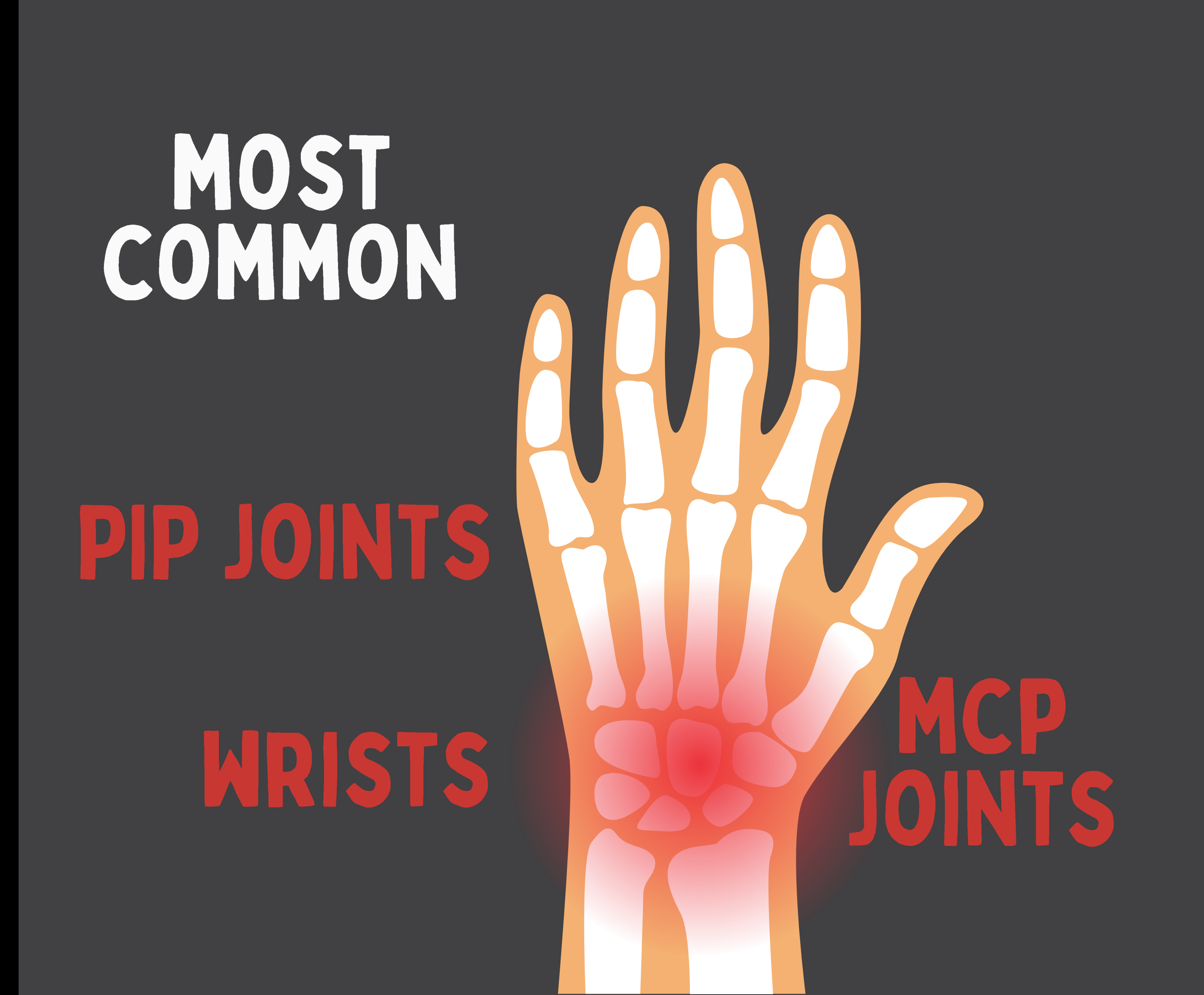
DISCUSS WITH RHEUMATOLOGY

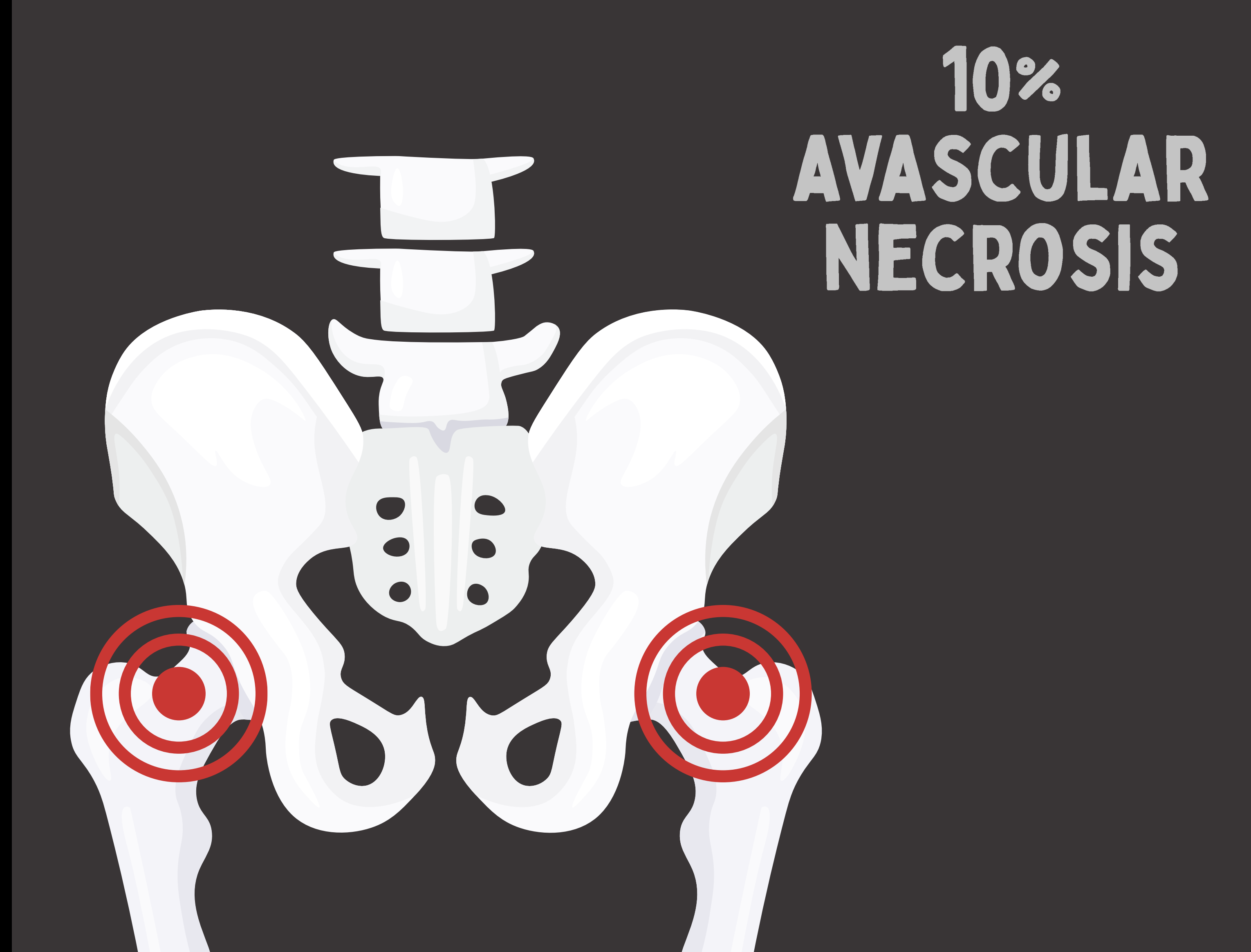
HYDROXYCHLOROQUINE

SHHHH...



90% OF PATIENTS WITH SLE WILL HAVE JOINT PAIN





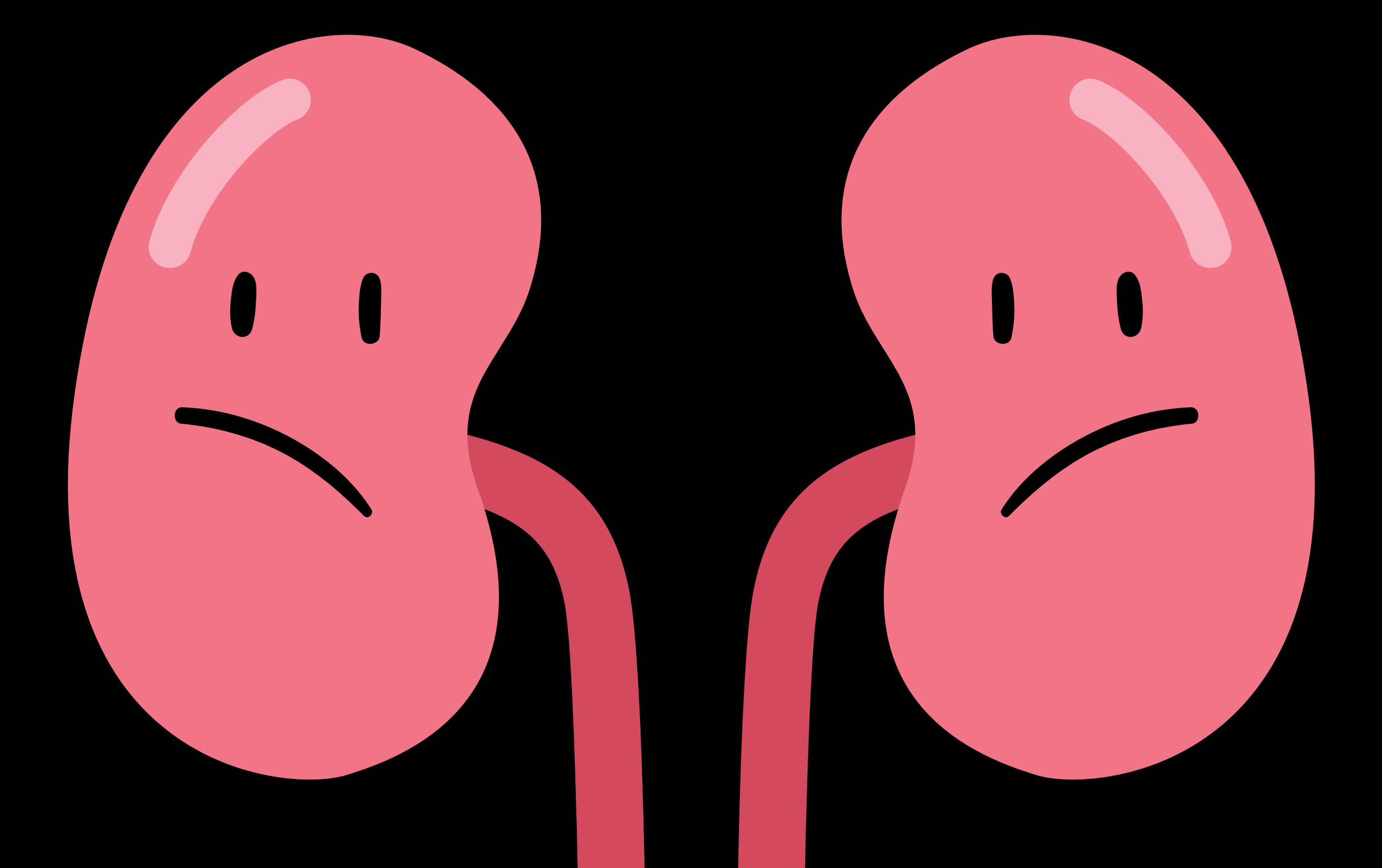
DRUG OF CHOICE

HCQ

NSAIDS

50% OF PATIENTS WITH SLE WILL HAVE RENAL DISEASE

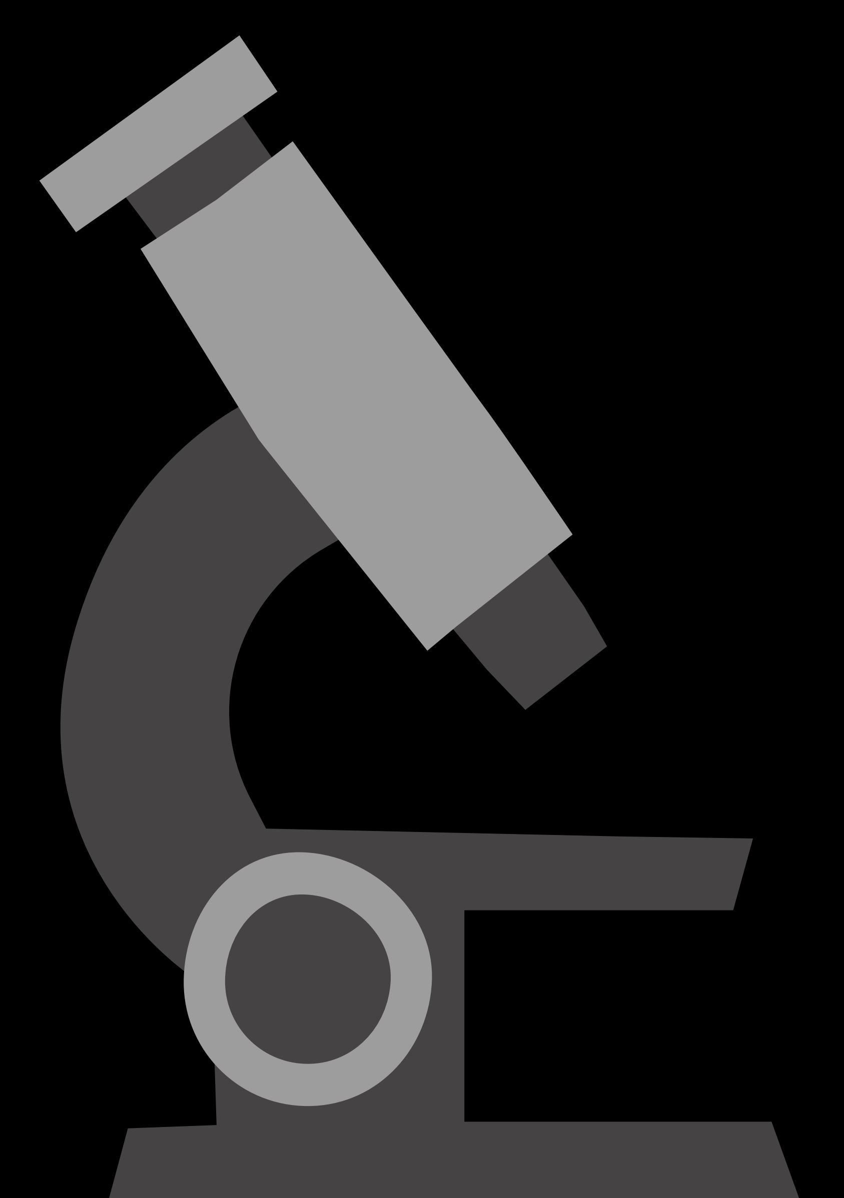
LUPUS NEPHRITIS



LUPUS NEPHRITIS

HYPERTENSION
EDEMA
HEMATURIA
NOCTURIA
POLYURIA
OLIGURIA
FOAMY URINE

LUPUS NEPHRITIS CAN LOOK LIKE NEPHRITIC OR NEPHROTIC SYNDROME



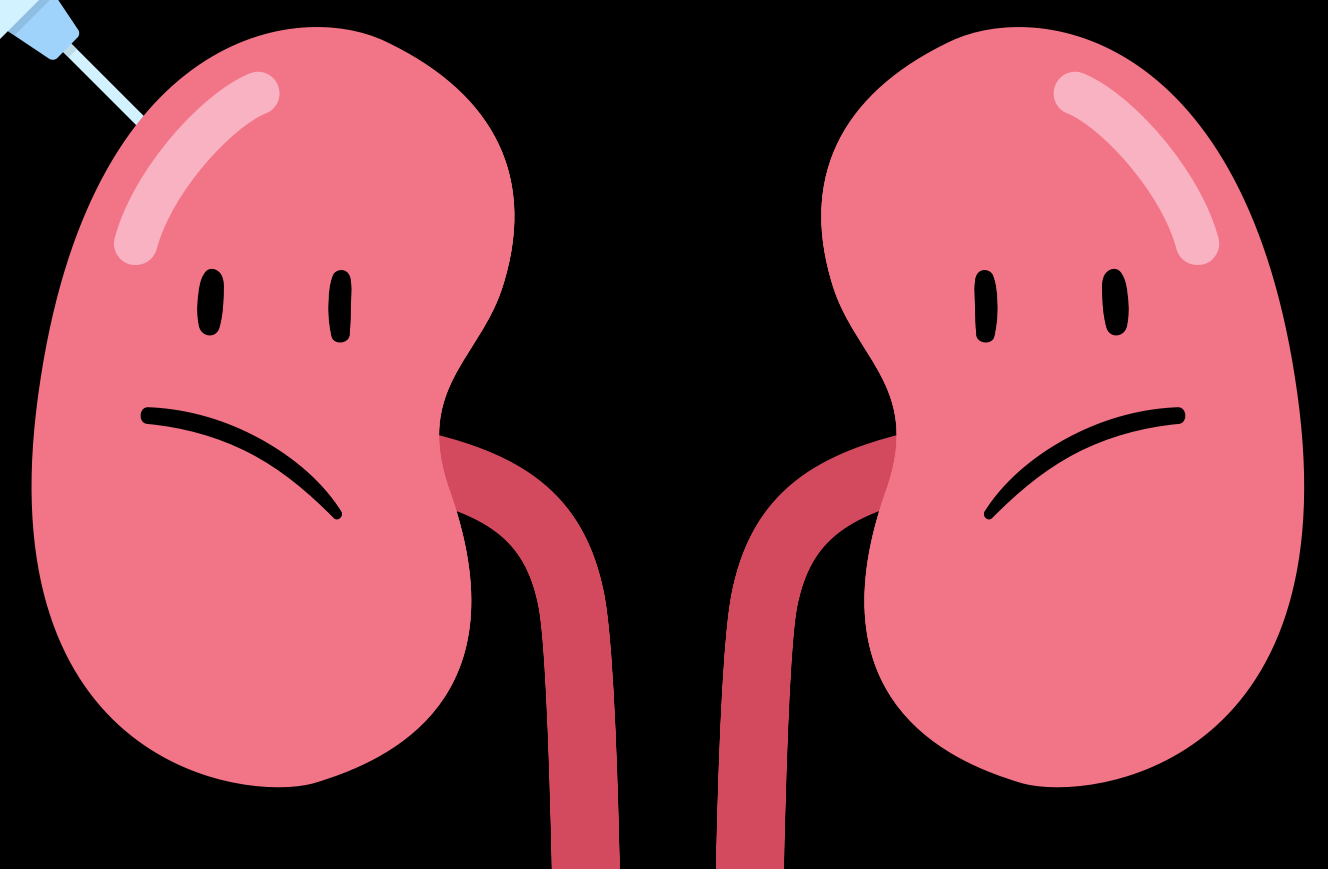
PROTEINURIA

HEMATURIA
GRANULAR CASTS

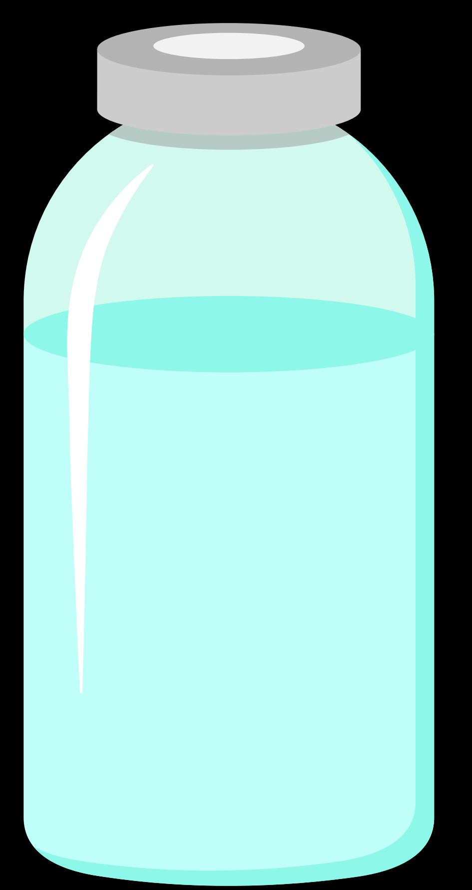
MAY NOT HAVE RENAL INSUFFICIENCY!

LUPUS NEPHRITIS

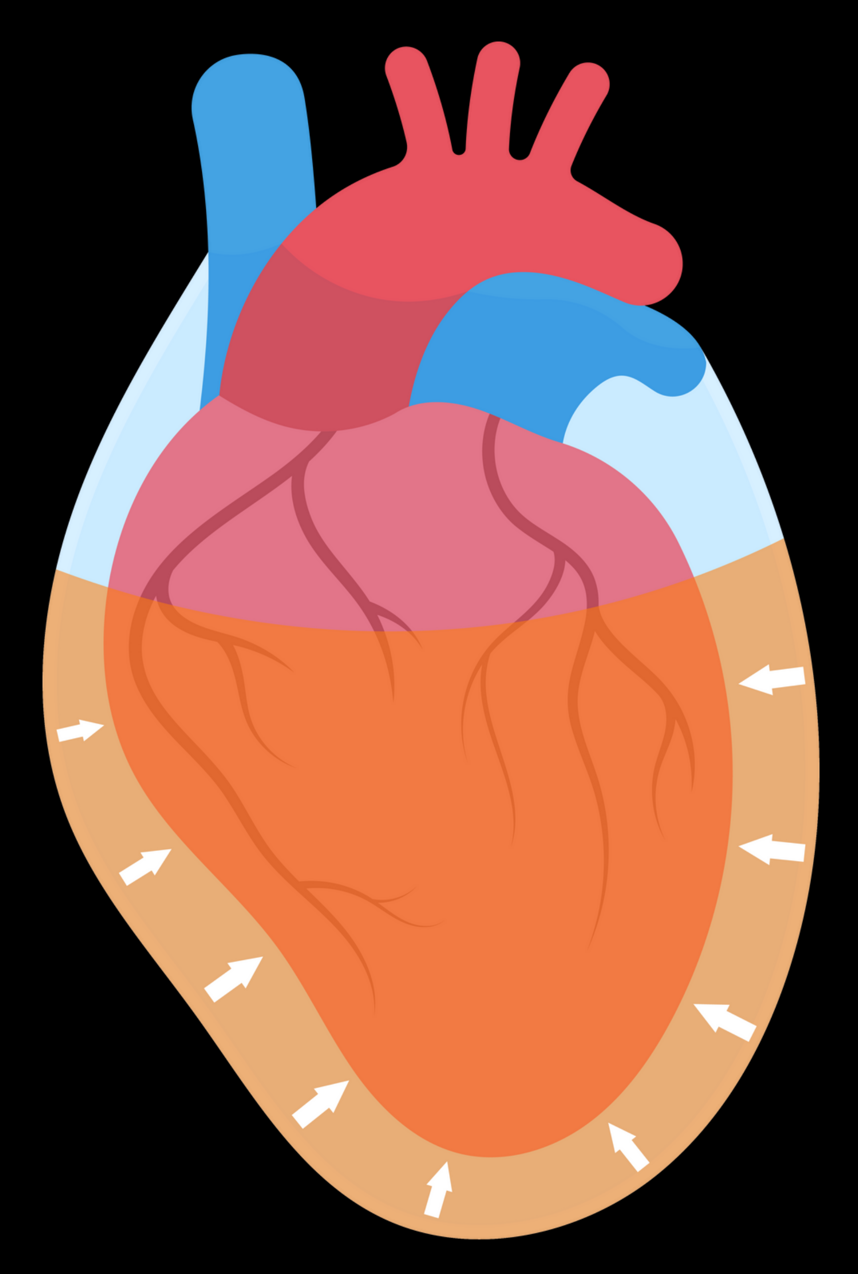
RENAL BIOPSY CONFIRMS THE DIAGNOSIS



HIGH-DOSE CORTICOSTEROIDS

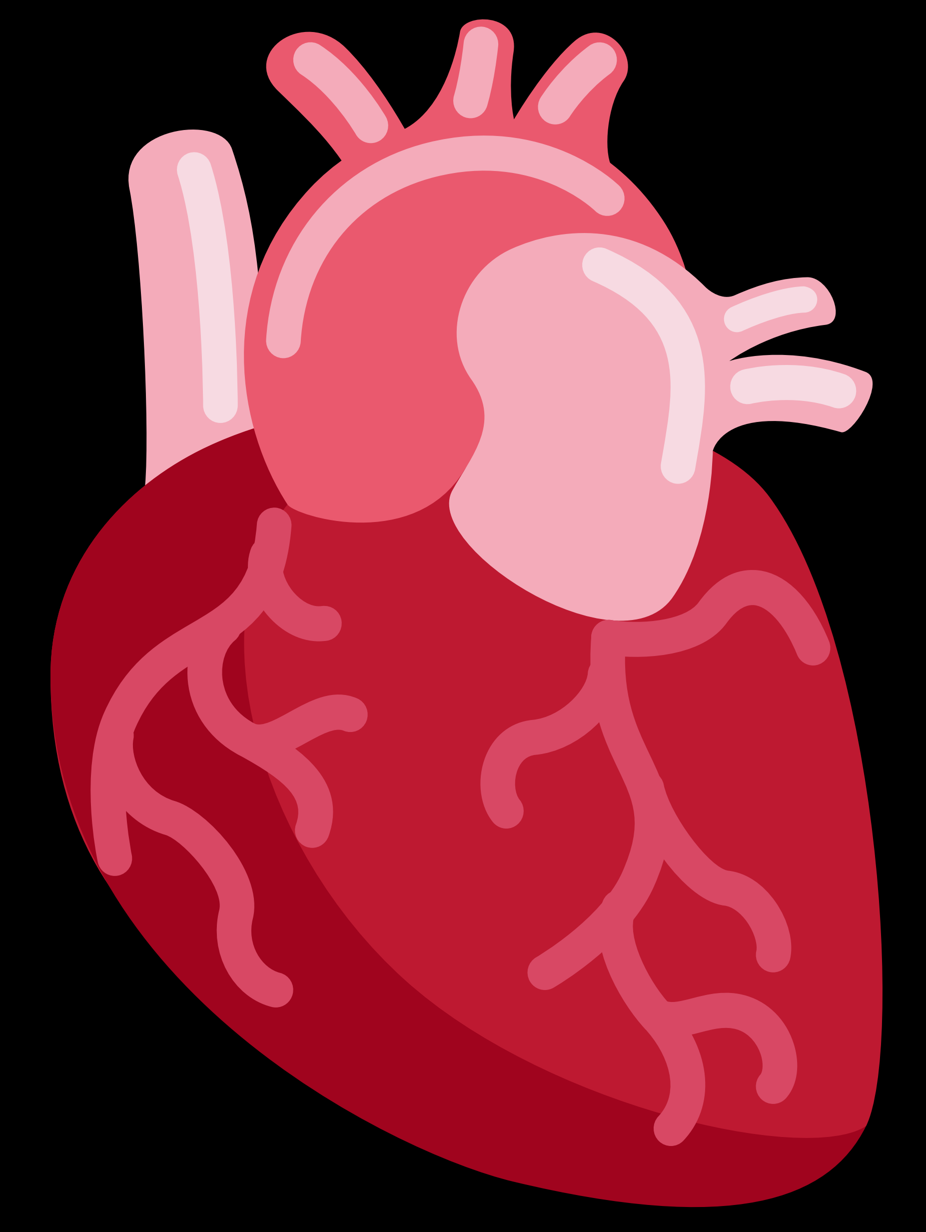


50% OF PATIENTS WITH SLE WILL HAVE CARDIAC DISEASE



BECAUSE SEROSITIS IS COMMON IN SLE

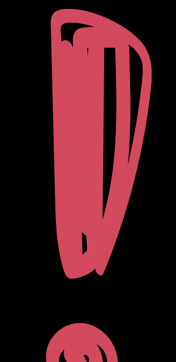
PERICARDITIS IS THE MOST COMMON CARDIAC PROBLEM

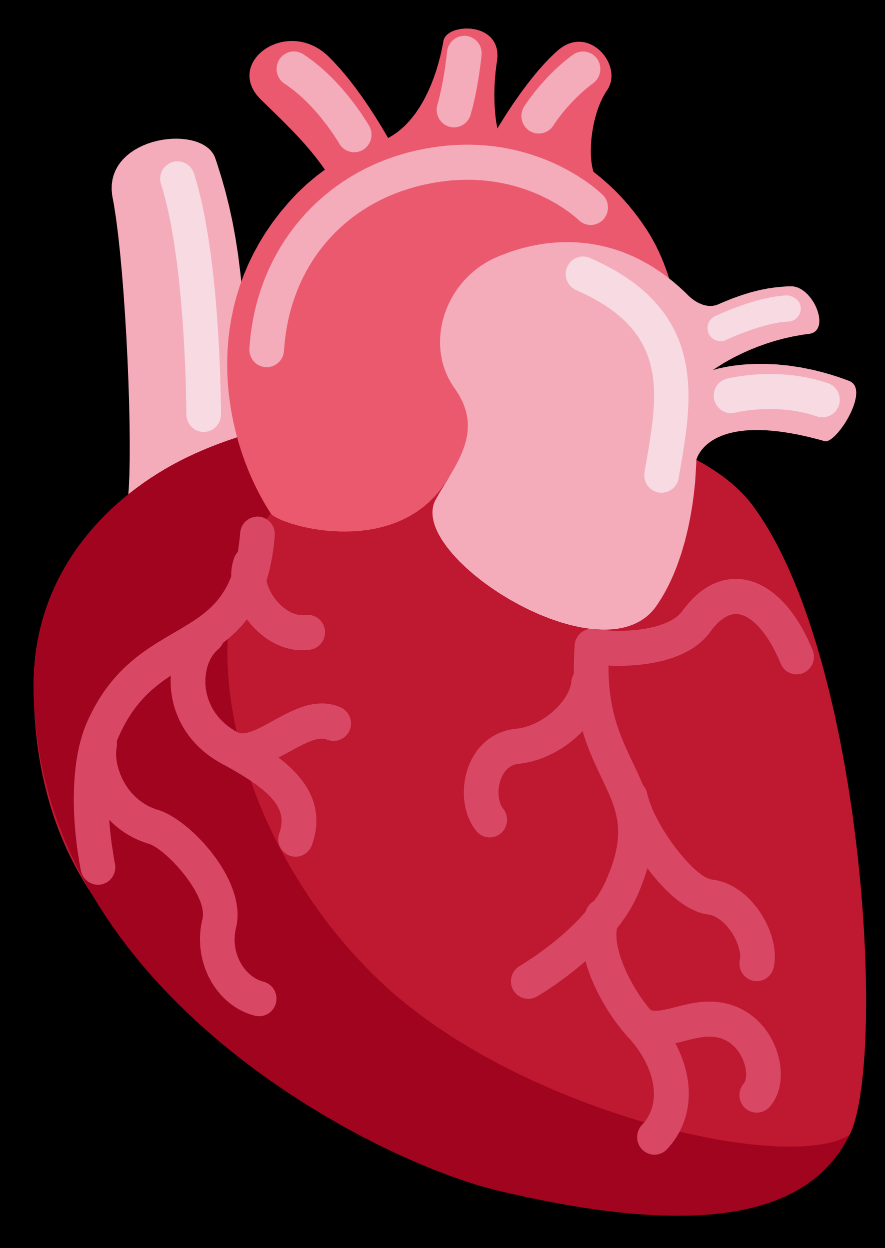


PATIENTS WITH SLE ARE AT **ESPECIALLY HIGH RISK** FOR CORONARY ARTERY DISEASE

CORONARY ARTERY VASCULITIS OR **GENERALIZED ATHEROSCLEROSIS**

YOUR 18- TO 39-YEAR-OLD LUPUS PATIENT HAS A HIGHER RISK OF HEART ATTACK AND STROKE THAN YOUR 50- TO 60-YEAR-OLD

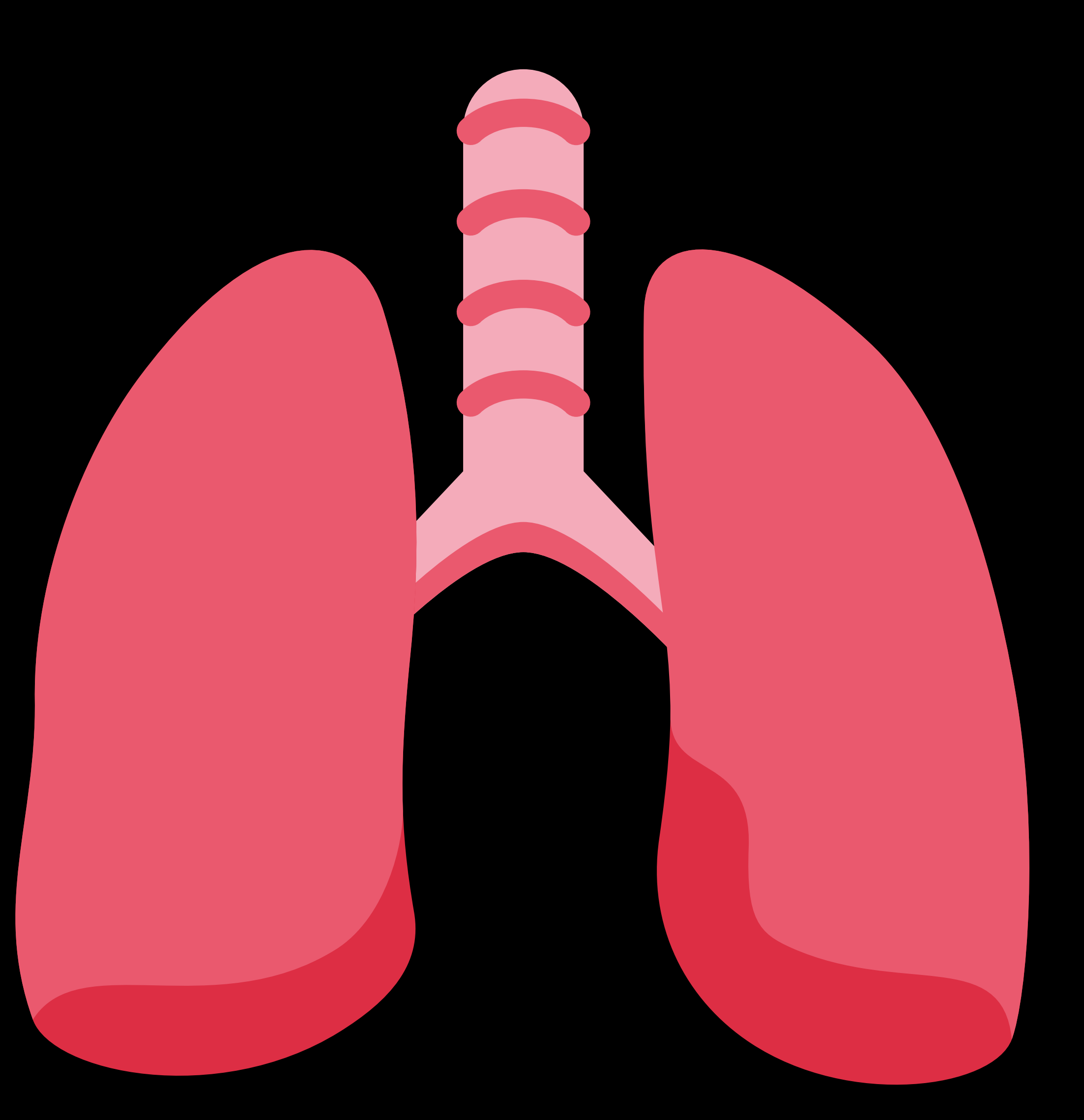




PERICARDITIS
CORONARY ARTERY DISEASE

LIBMAN-SACKS ENDOCARDITIS
MYOCARDITIS
CORONARY ARTERITIS
HYPERTENSION

AT LEAST 50% OF PATIENTS WITH SLE WILL HAVE PULMONARY DISEASE



BECAUSE SEROSITIS IS COMMON IN SLE

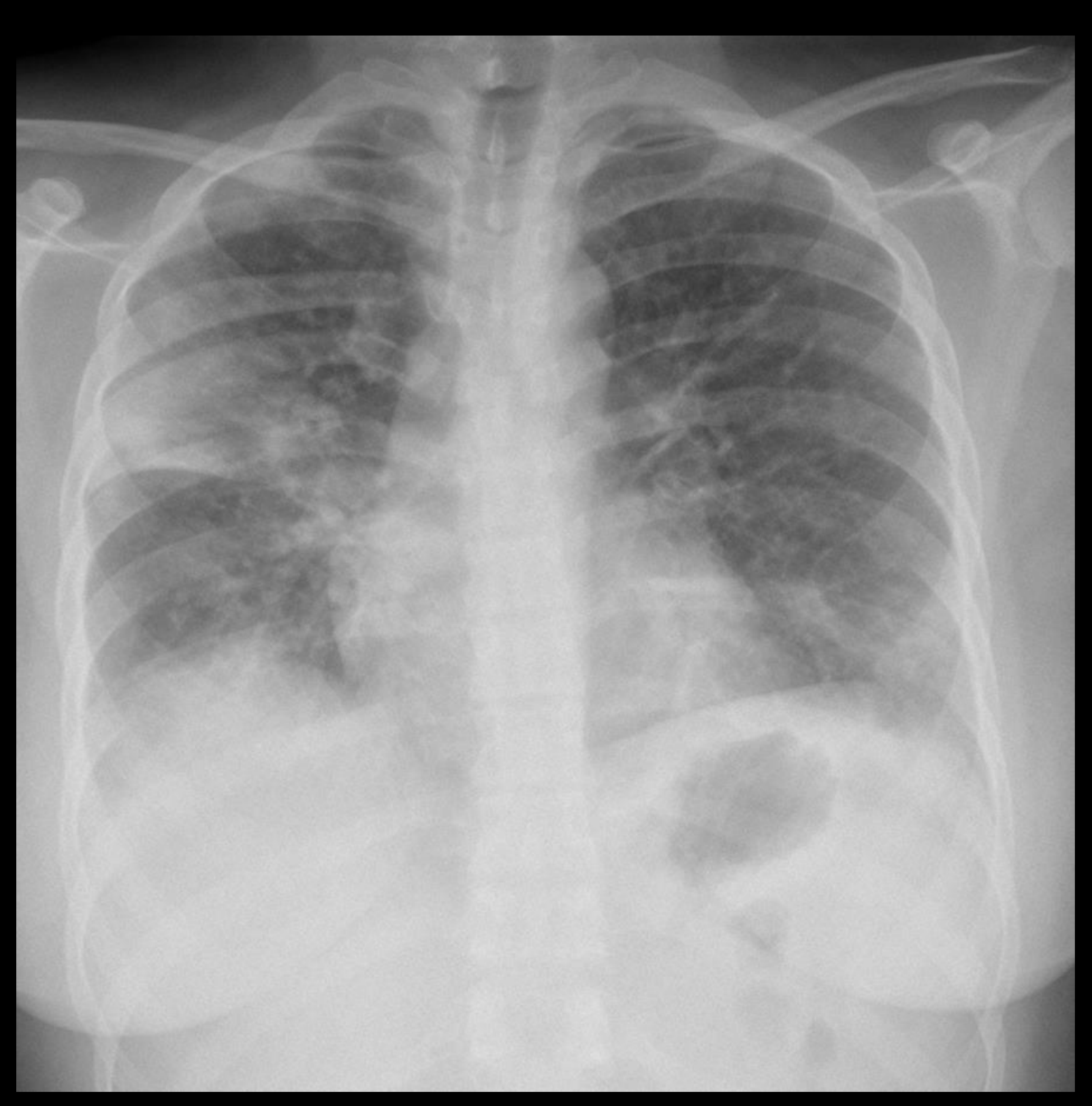
PLEURITIS IS THE MOST COMMON PULMONARY PROBLEM

ACUTE LUPUS PNEUMONITIS

AFFECTS ABOUT 4% OF PATIENTS WITH SLE
MORTALITY 50%

SYMPTOMS

FEVER
COUGH
SOB
PLEURITIC PAIN



DIAGNOSIS

LUNG BIOPSY TO DISTINGUISH FROM INFECTION

MANAGEMENT

HIGH-DOSE IV PULSE CORTICOSTEROIDS

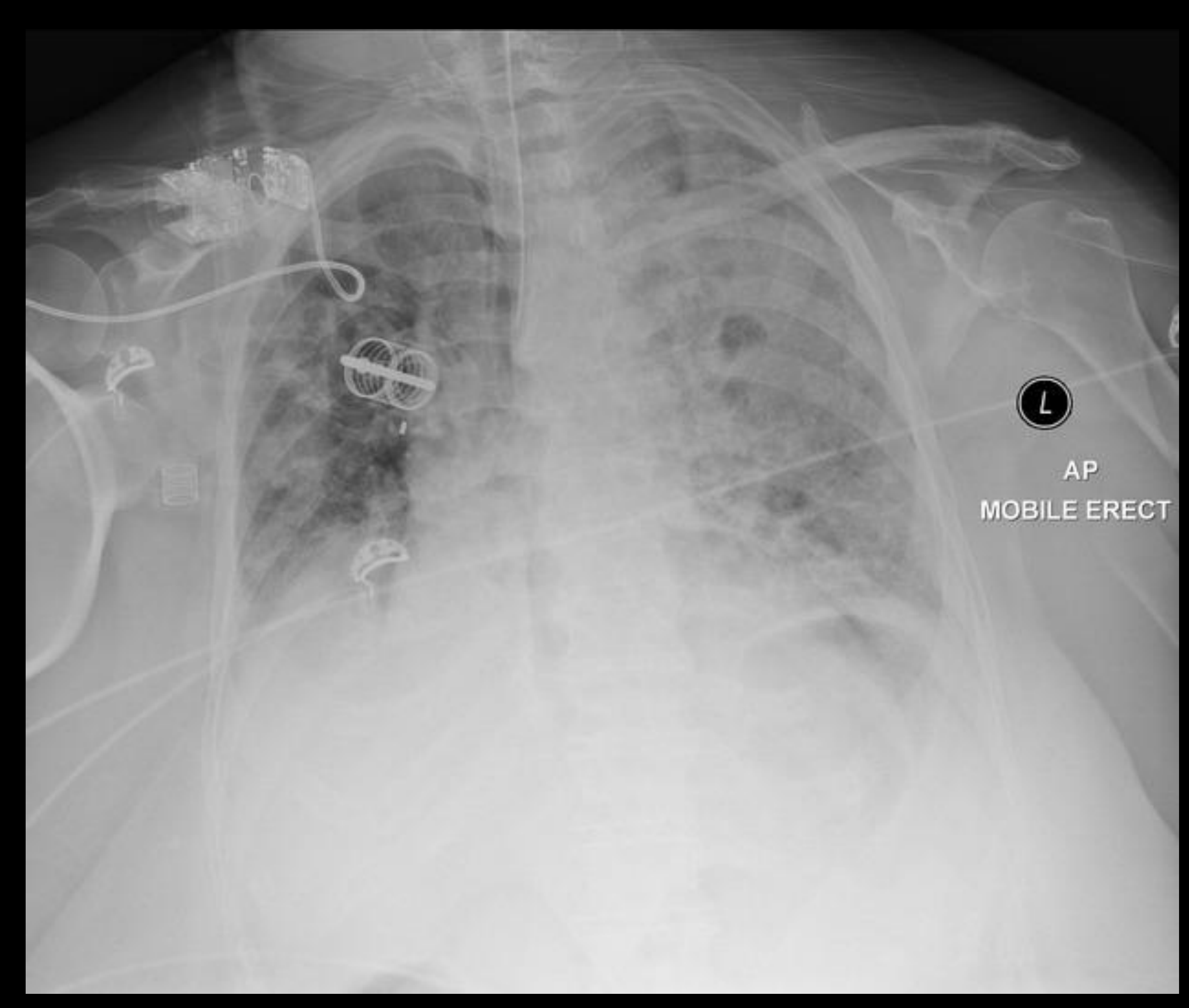
ALVEOLAR HEMORRHAGE

AFFECTS <2% OF PATIENTS WITH SLE

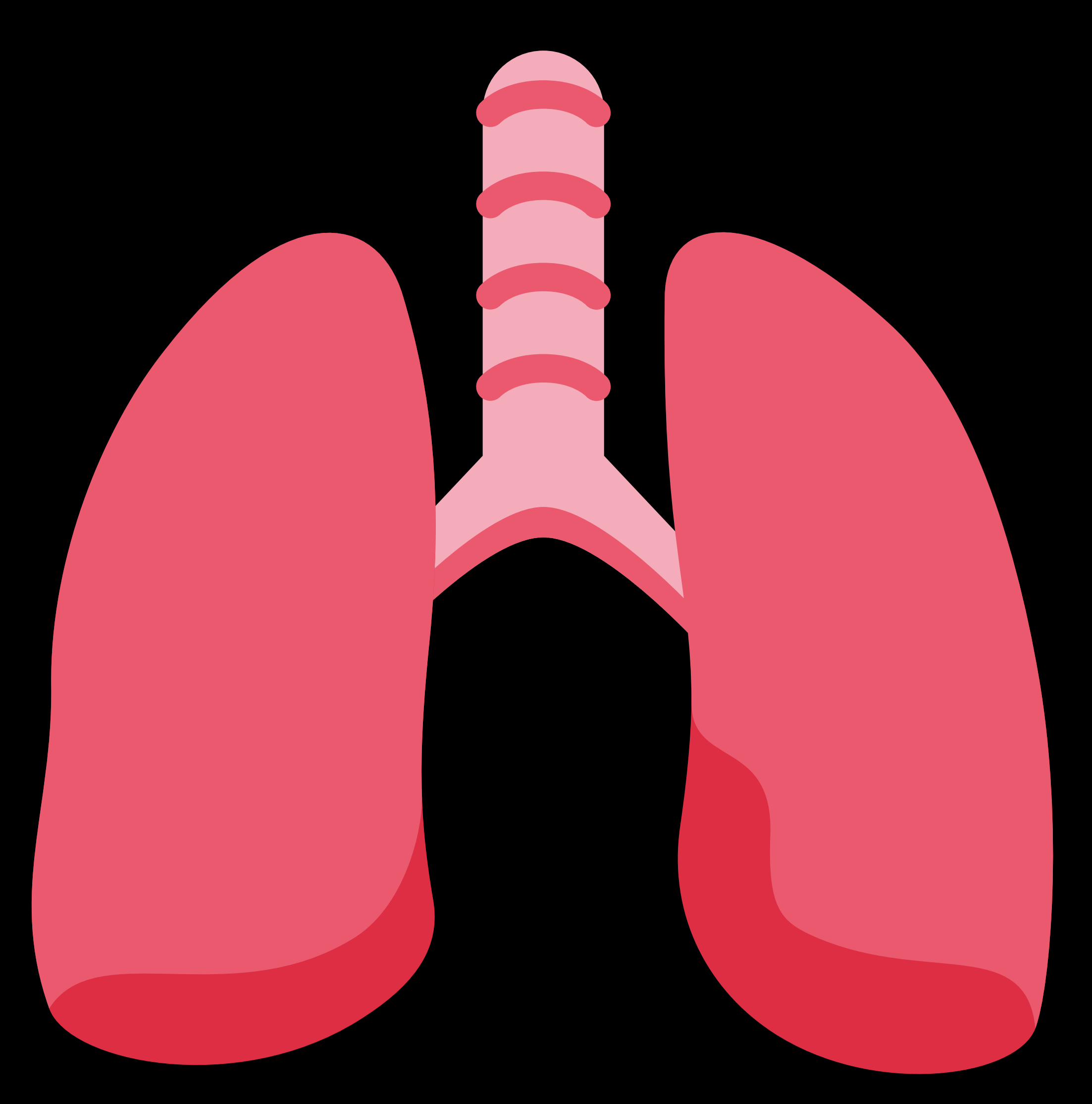
HIGH MORTALITY (MOST ESTIMATE 85%)

OCCURS IN THE SETTING OF PULMONARY VASCULITIS

REQUIRES PROMPT IMMUNOSUPPRESSION AND PLASMAPHERESIS



OTHER PULMONARY COMPLICATIONS OF LUPUS



PNEUMONIA/PNEUMONITIS
PLEURAL EFFUSIONS
PULMONARY EMBOLISM
SHRINKING LUNG SYNDROME
INTERSTITIAL LUNG DISEASE
PULMONARY HYPERTENSION

EARLY ATHEROSCLEROSIS

IMMUNO SUPPRESSION

HYPERCOAGULABLE STATE

VASCULITIS

AUTO ANTIBODIES

NEUROLOGIC INVOLVEMENT

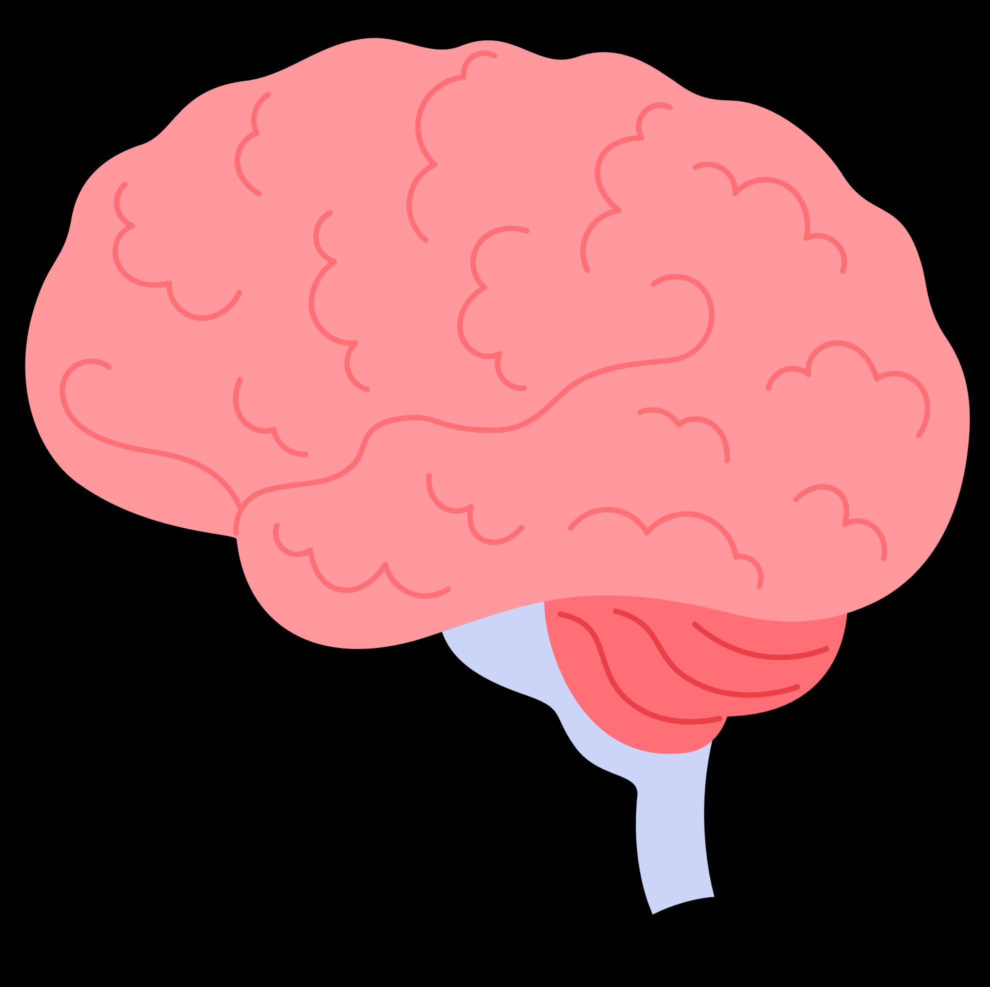
IN 75% OF PEOPLE WITH SLE

MULTIFOCAL MICROINFARCTS
CORTICAL ATROPHY
HEMORRHAGE
ISCHEMIC DEMYELINATION

CHRONIC, DIFFUSE CEREBRAL ISCHEMIA

NEUROLOGIC INVOLVEMENT

- STROKE/TIA
- CENTRAL VENOUS SINUS THROMBOSIS
- COGNITIVE DISORDERS
- INTRACTABLE HEADACHES
- DEMYELINATING DISORDERS
- MOVEMENT DISORDERS
- PSYCHIATRIC DISORDERS
- SEIZURE DISORDERS
- TRANSVERSE MYELOPATHY
- AUTONOMIC NEUROPATHY
- PERIPHERAL NEUROPATHY
- SENSORINEURAL HEARING LOSS
- MYASTHENIA GRAVIS
- CRANIAL NEUROPATHY



ANY NEUROLOGIC OR PSYCHIATRIC SYMPTOMS WARRANT CONSIDERATION OF LUPUS AS THE CAUSE

PSYCHOSIS

TREATMENT

- IMMUNOSUPPRESSION
- HIGH-DOSE CORTICOSTEROIDS
- LIFELONG ANTICOAGULATION

HEMATOLOGIC INVOLVEMENT

- ANEMIA
- LEUKOPENIA
- THROMBOCYTOPENIA
- AUTOIMMUNE HEMOLYTIC ANEMIA
- IDIOPATHIC THROMBOCYTOPENIC PURPURA
- THROMBOTIC THROMBOCYTOPENIC PURPURA

40% OF PEOPLE WITH SLE ALSO HAVE ANTIPHOSPHOLIPID SYNDROME

IT'S HARD TO DETERMINE WHETHER SYMPTOMS ARE INFLAMMATORY OR EMBOLIC

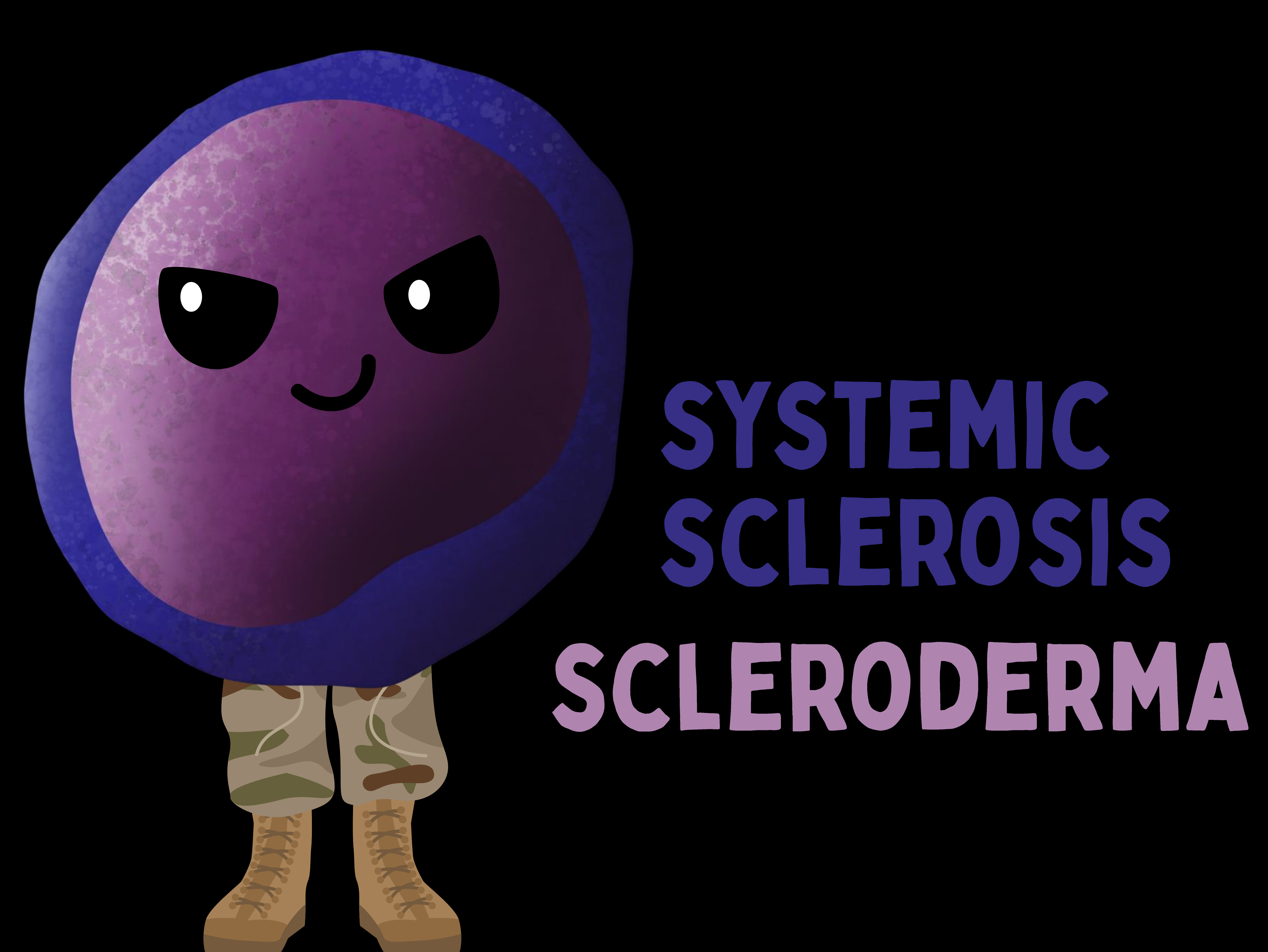
LUPUS IN PREGNANCY IS HIGH RISK!

FOR MOM

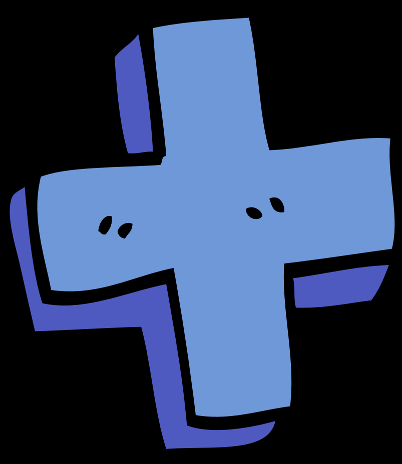
- VENOUS THROMBOEMBOLISM
- THROMBOCYTOPENIA
- PREGNANCY LUPUS FLARES
- PREECLAMPSIA
- UNPLANNED C-SECTION
- INFECTION
- POSTPARTUM LUPUS FLARES
- 5X HIGHER MORTALITY

FOR BABY

- SPONTANEOUS ABORTION
- PLACENTAL INSUFFICIENCY
- PLACENTAL THROMBOSIS
- INTRAUTERINE FETAL DEMISE
- INTRAUTERINE GROWTH RESTRICTION
- PREMATURE BIRTH
- NEONATAL LUPUS

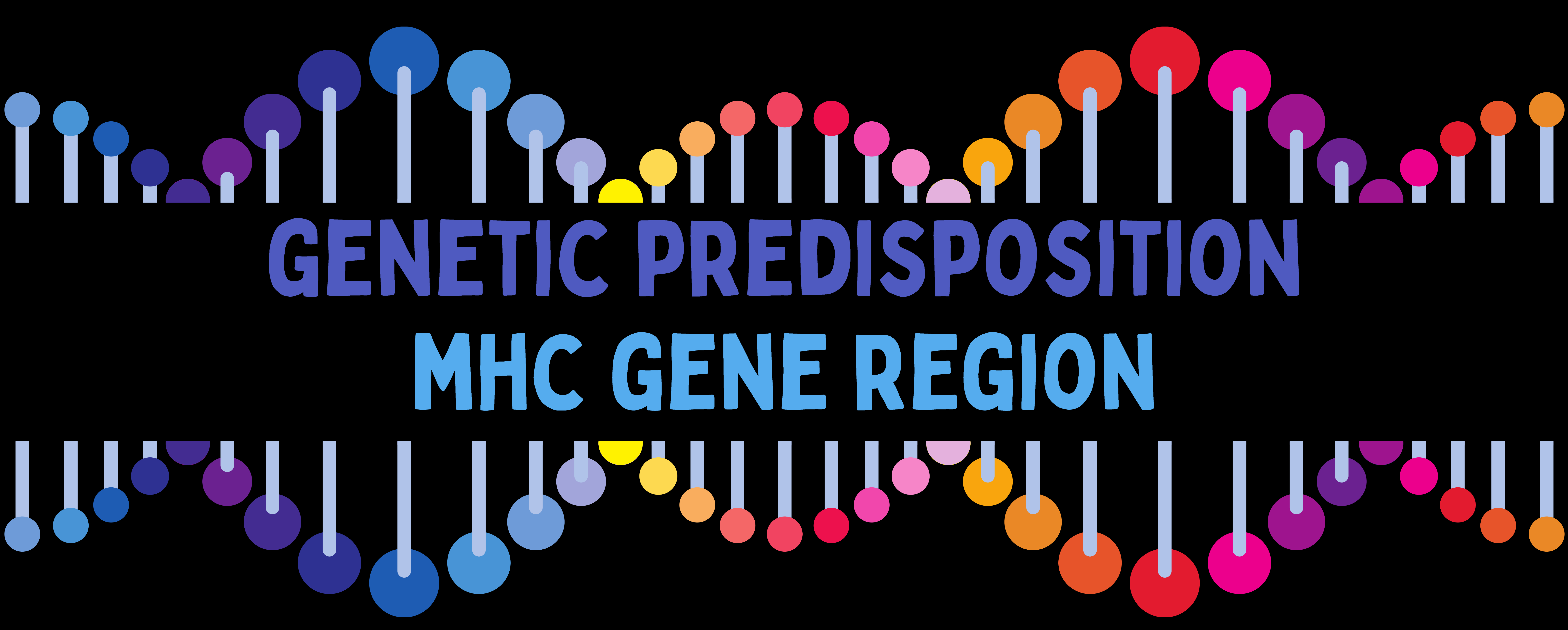

SYSTEMIC
SCLEROSIS
SCLERODERMA

GENETIC
PREDISPOSITION



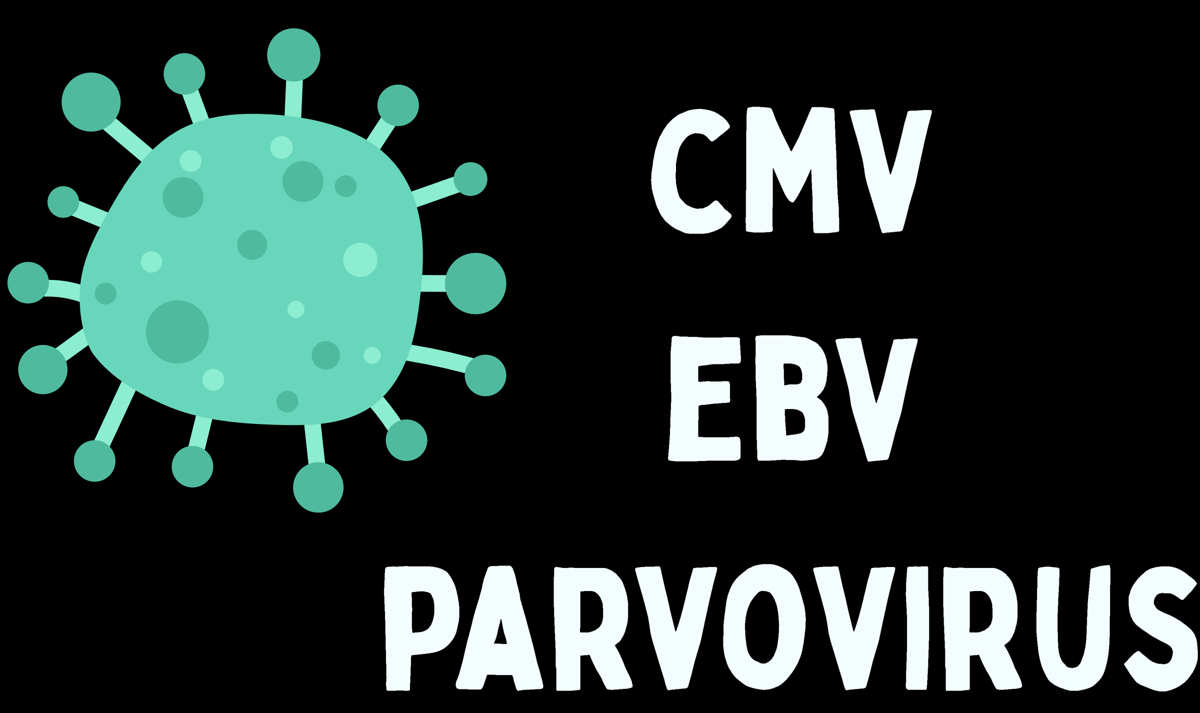
TRIGGER

INFECTION OR OTHER
ENVIRONMENTAL TRIGGER


GENETIC PREDISPOSITION
MHC GENE REGION

TRIGGERS

INFECTIOUS



CMV

EBV

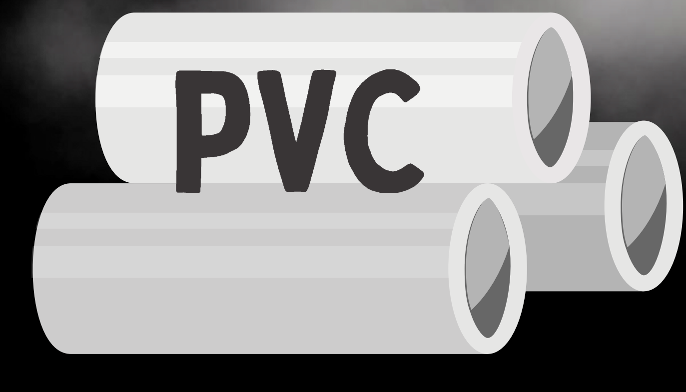
PARVOVIRUS

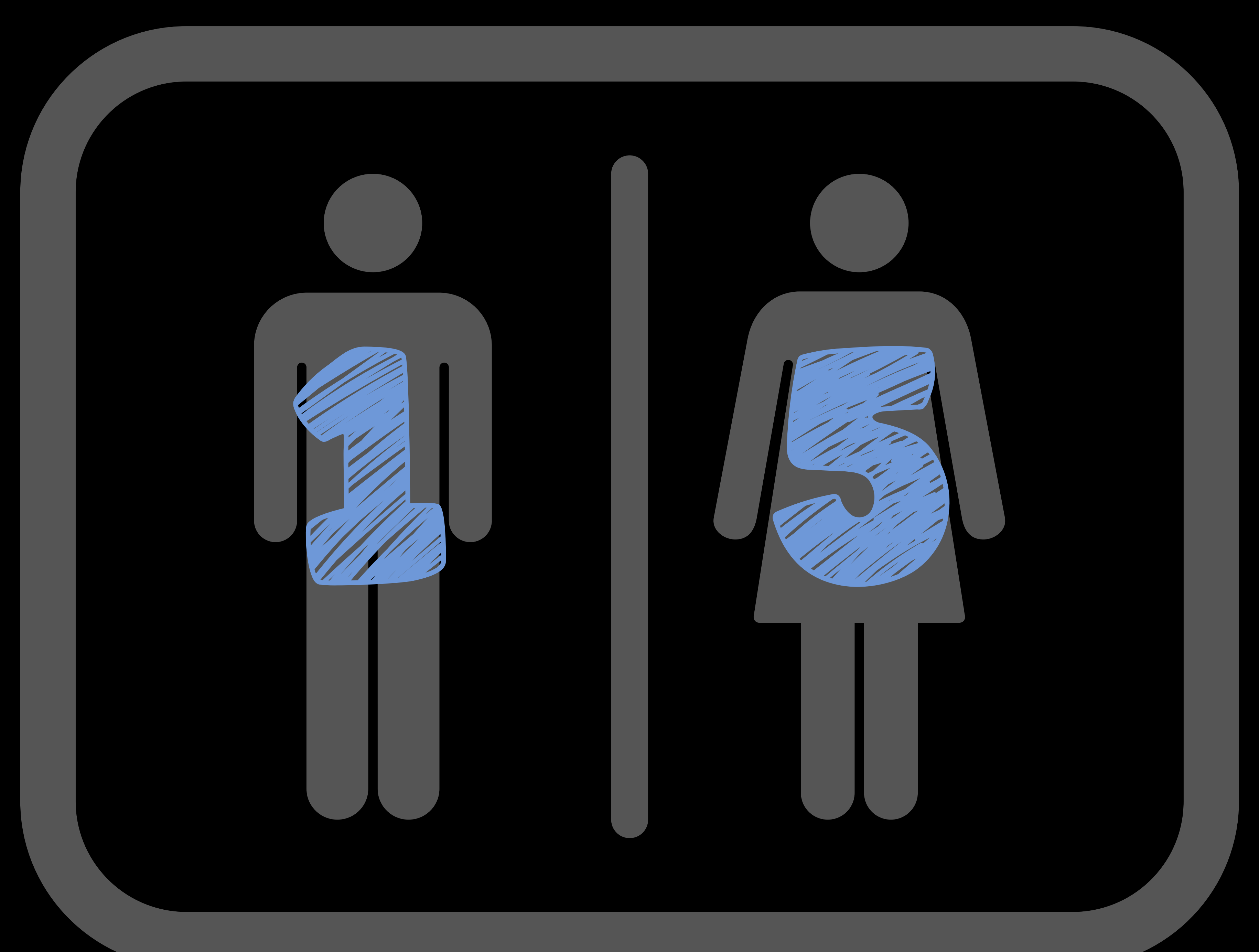
ENVIRONMENTAL

SILICA DUST

TOLUENE

XYLENE

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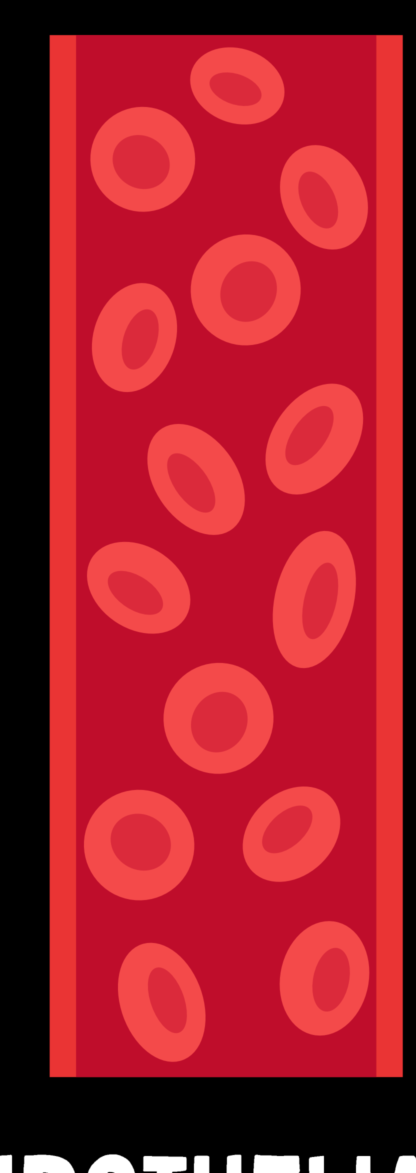
5

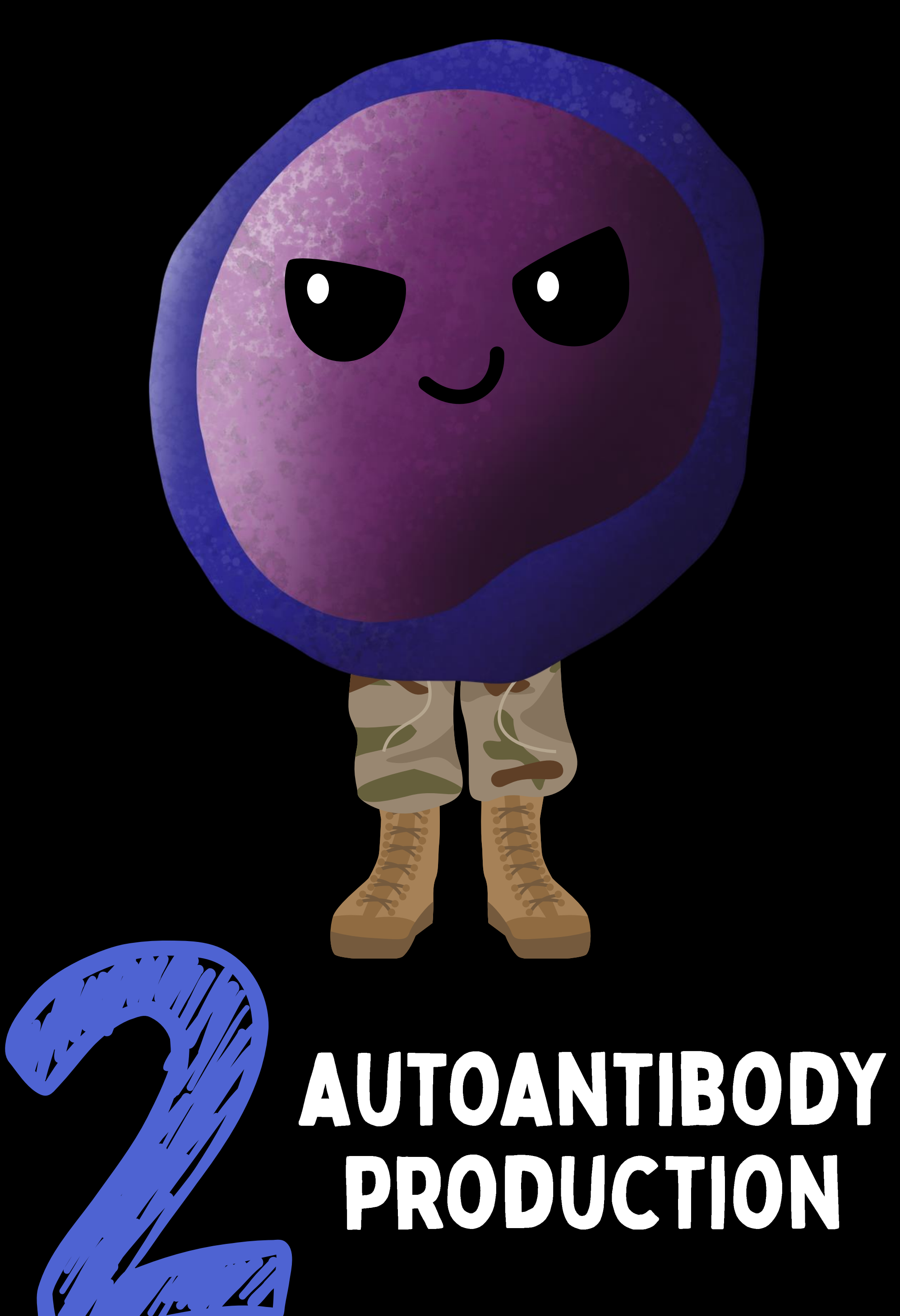
AVERAGE AGE AT ONSET

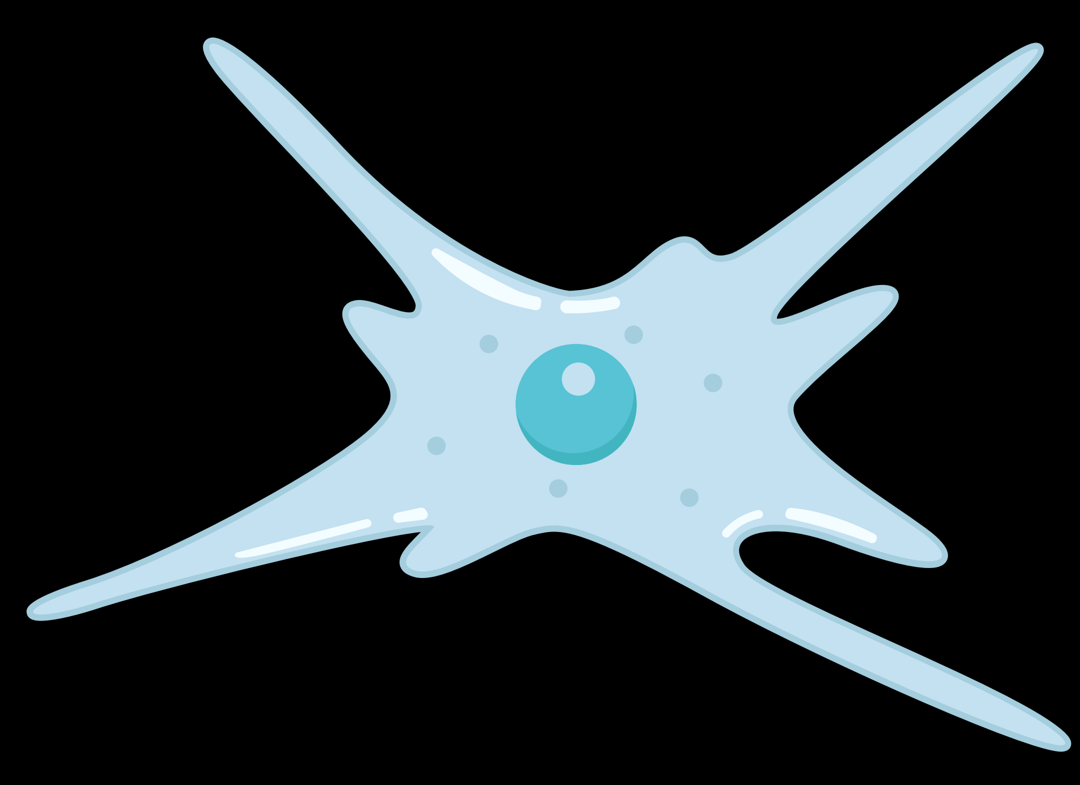
45-65 YEARS

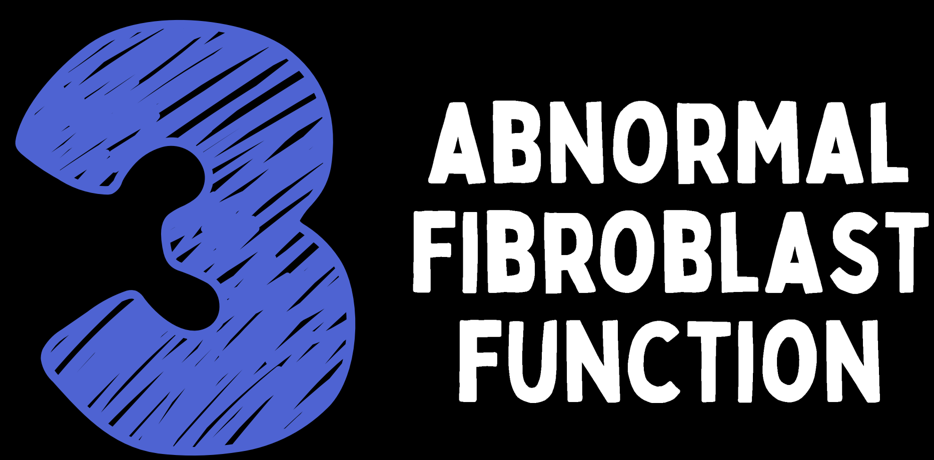
SYSTEMIC SCLEROSIS



1 ENDOTHELIAL CELL DYSFUNCTION







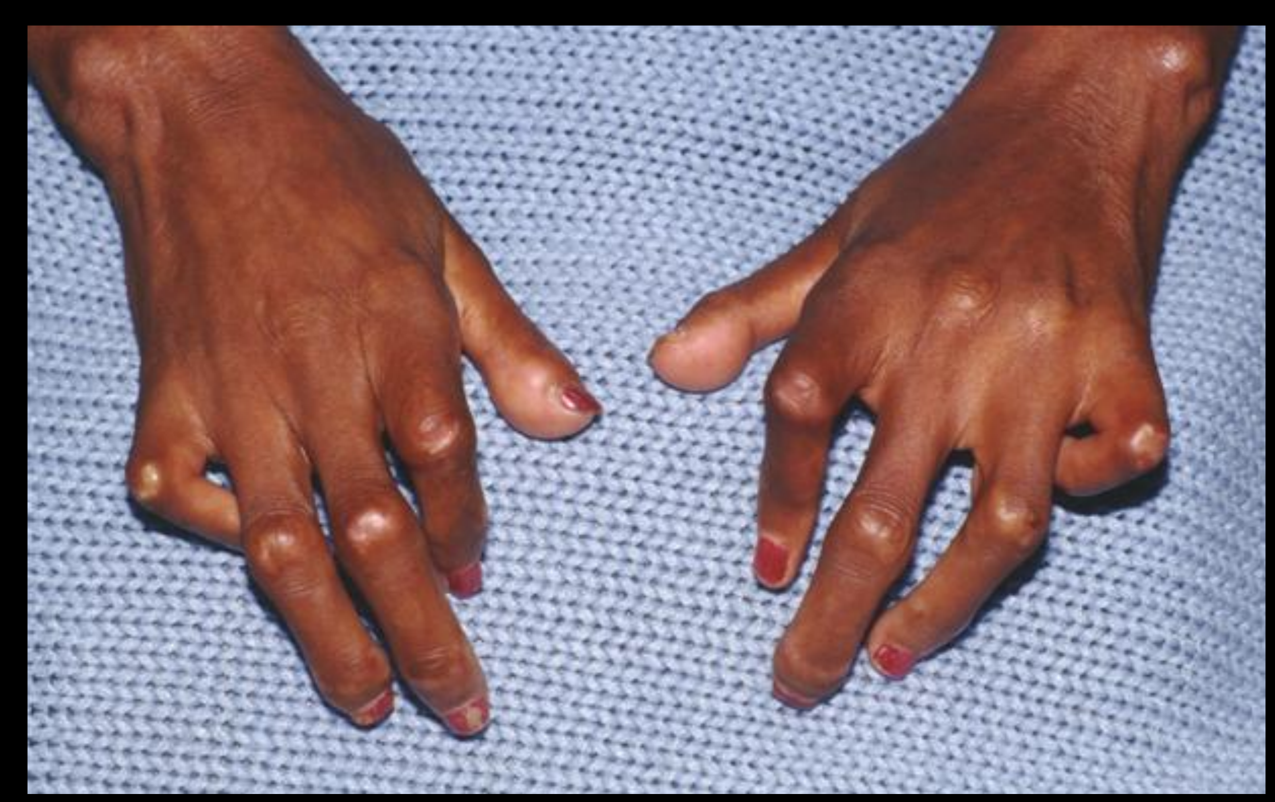
OVERACTIVE FIBROBLASTS

DEPOSIT COLLAGEN AND OTHER EXTRACELLLULAR MATRIX MACROMOLECULES IN THE SKIN AND VISCERAL ORGANS

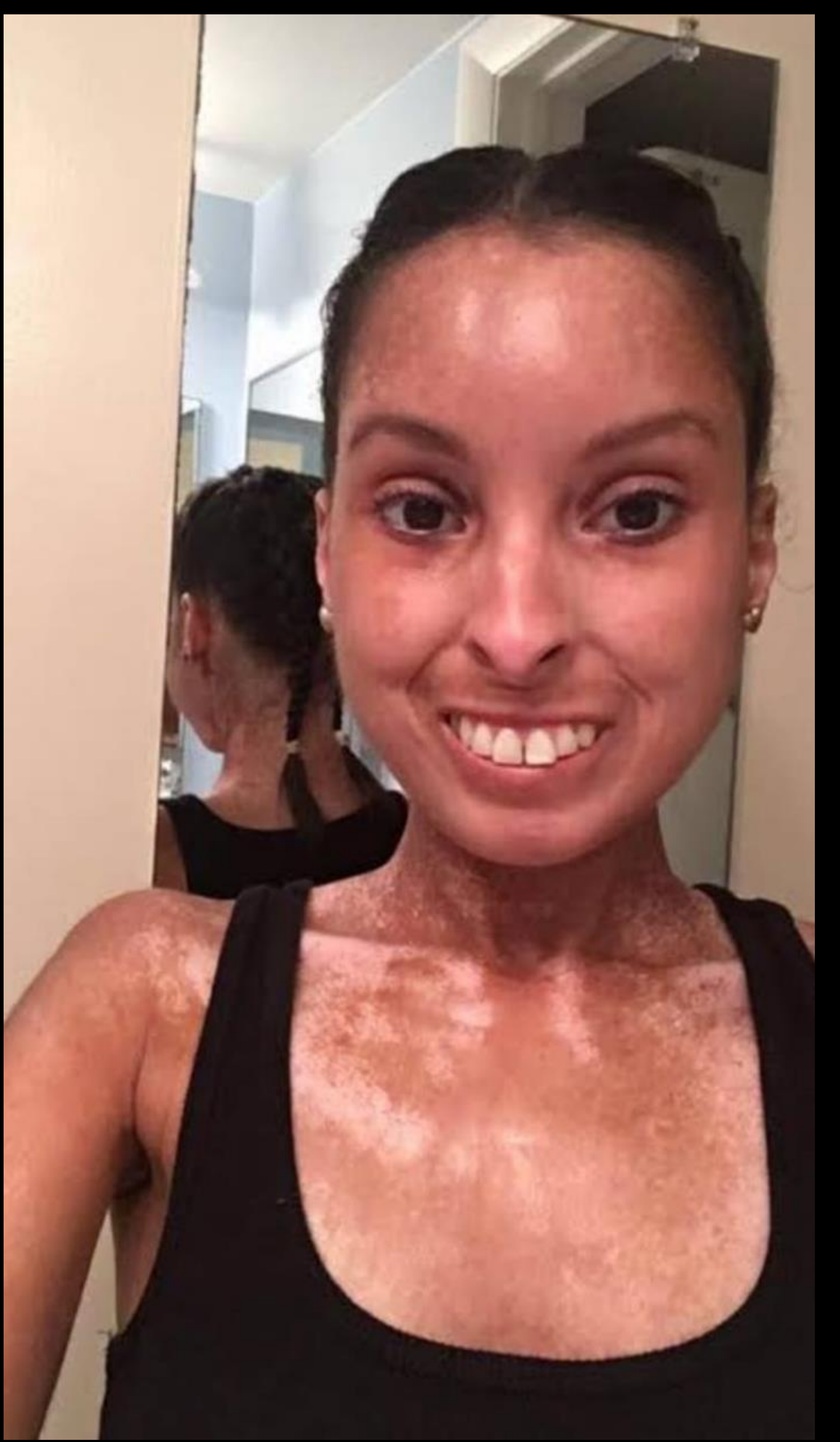
CATEGORIZED BY THE PATTERN OF SKIN INVOLVEMENT

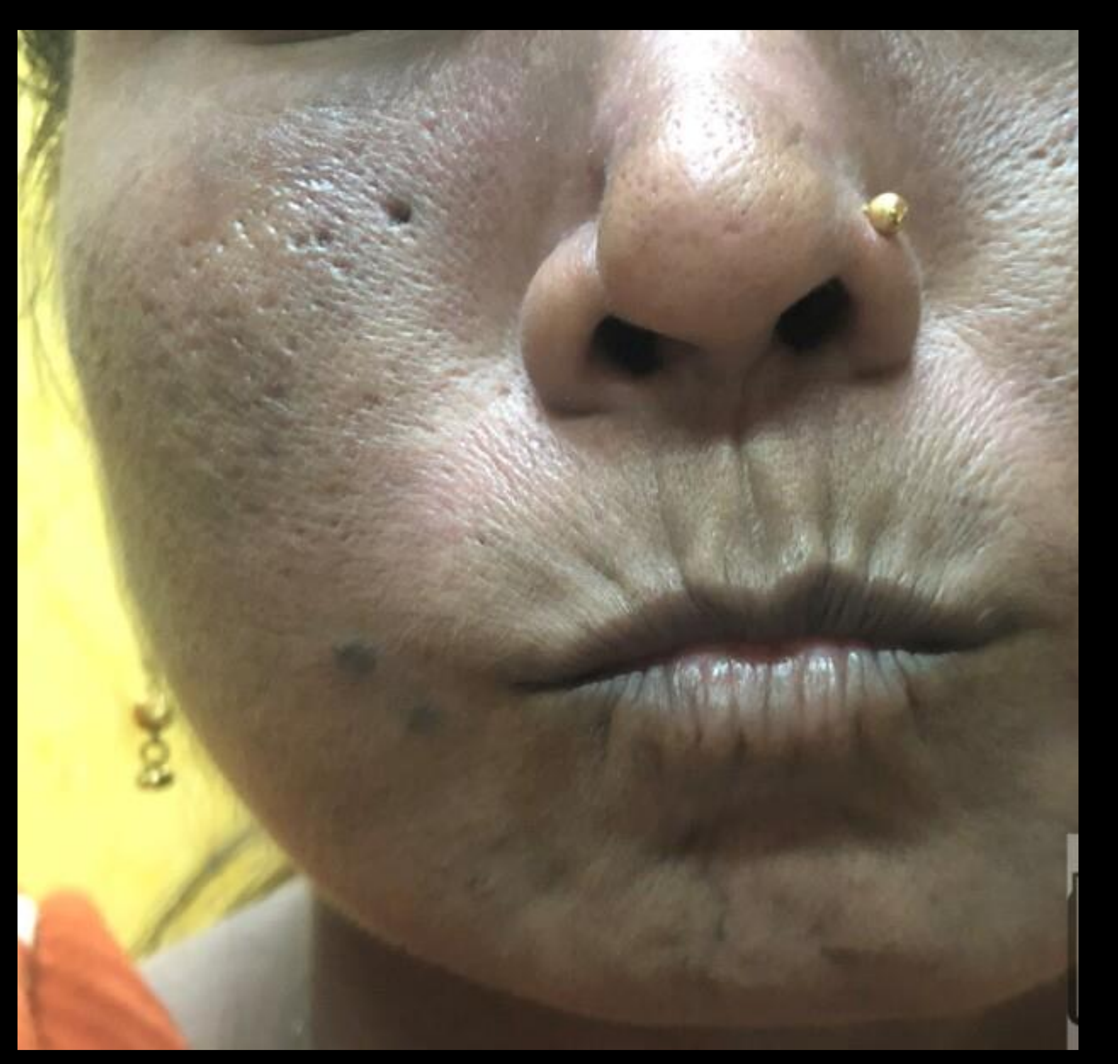
DIFFUSE DCSSC

SKIN FINDINGS EXTEND PROXIMAL AND INVOLVE THE TRUNK

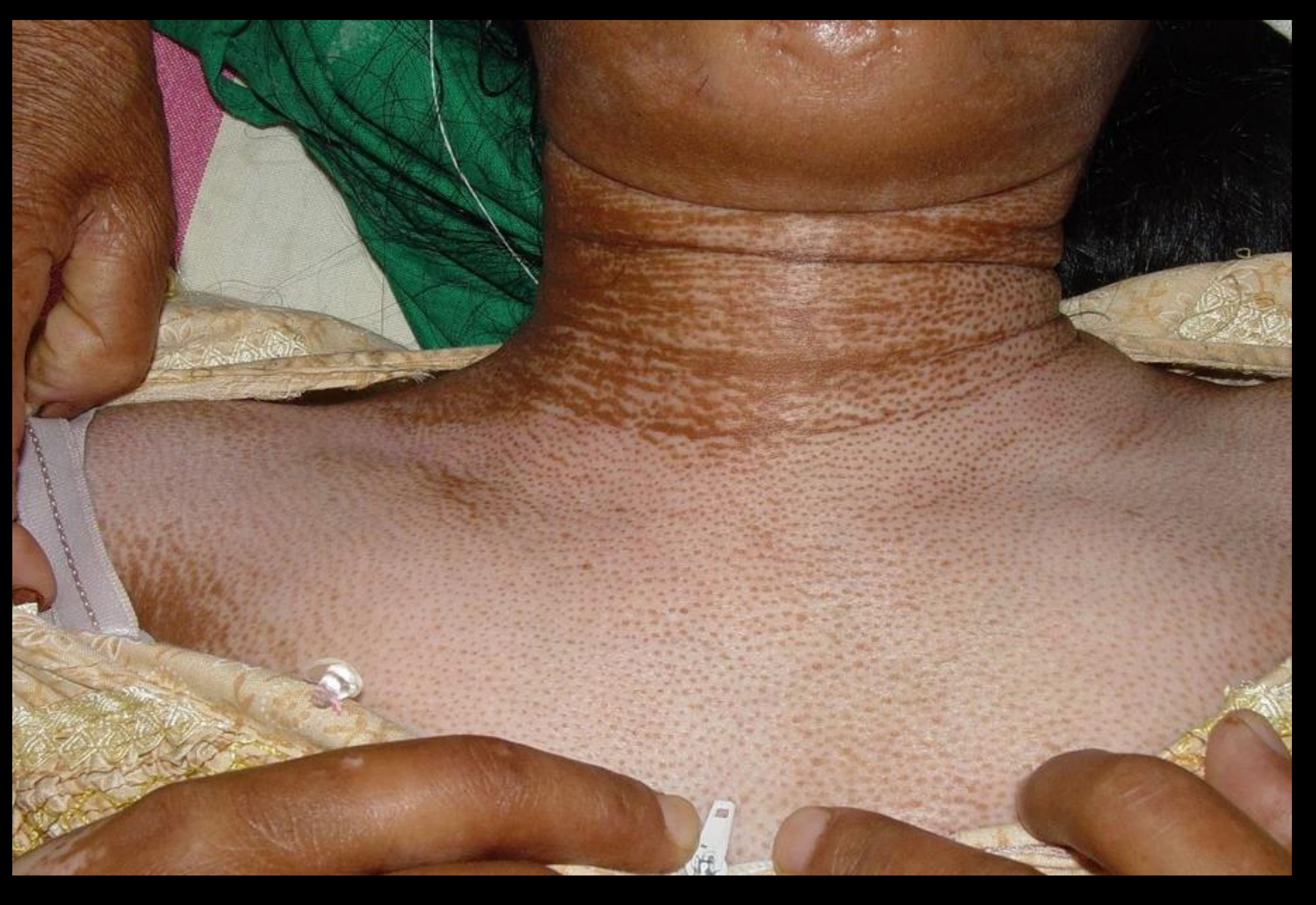


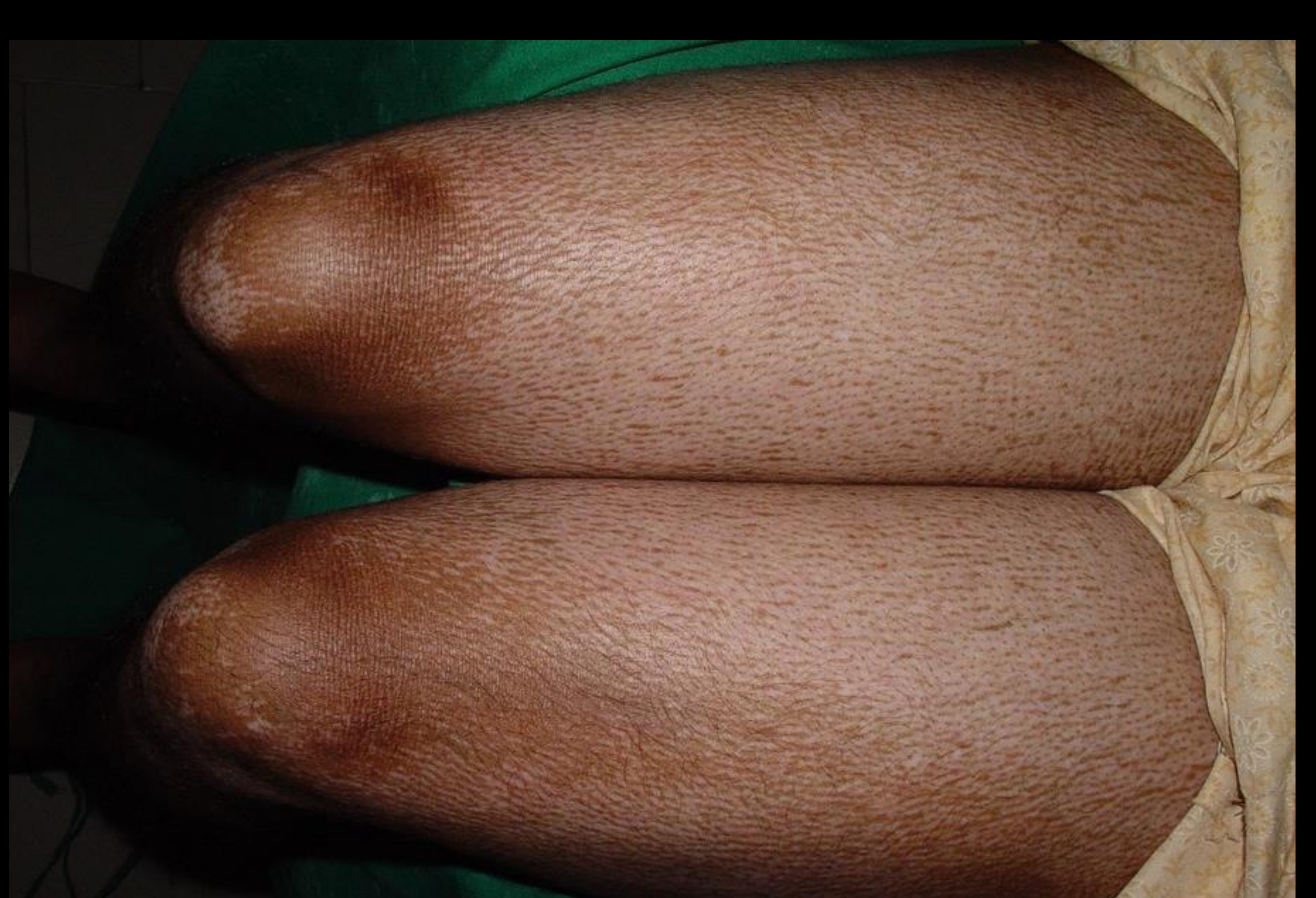
SCLERODACTYLY





FISH MOUTH

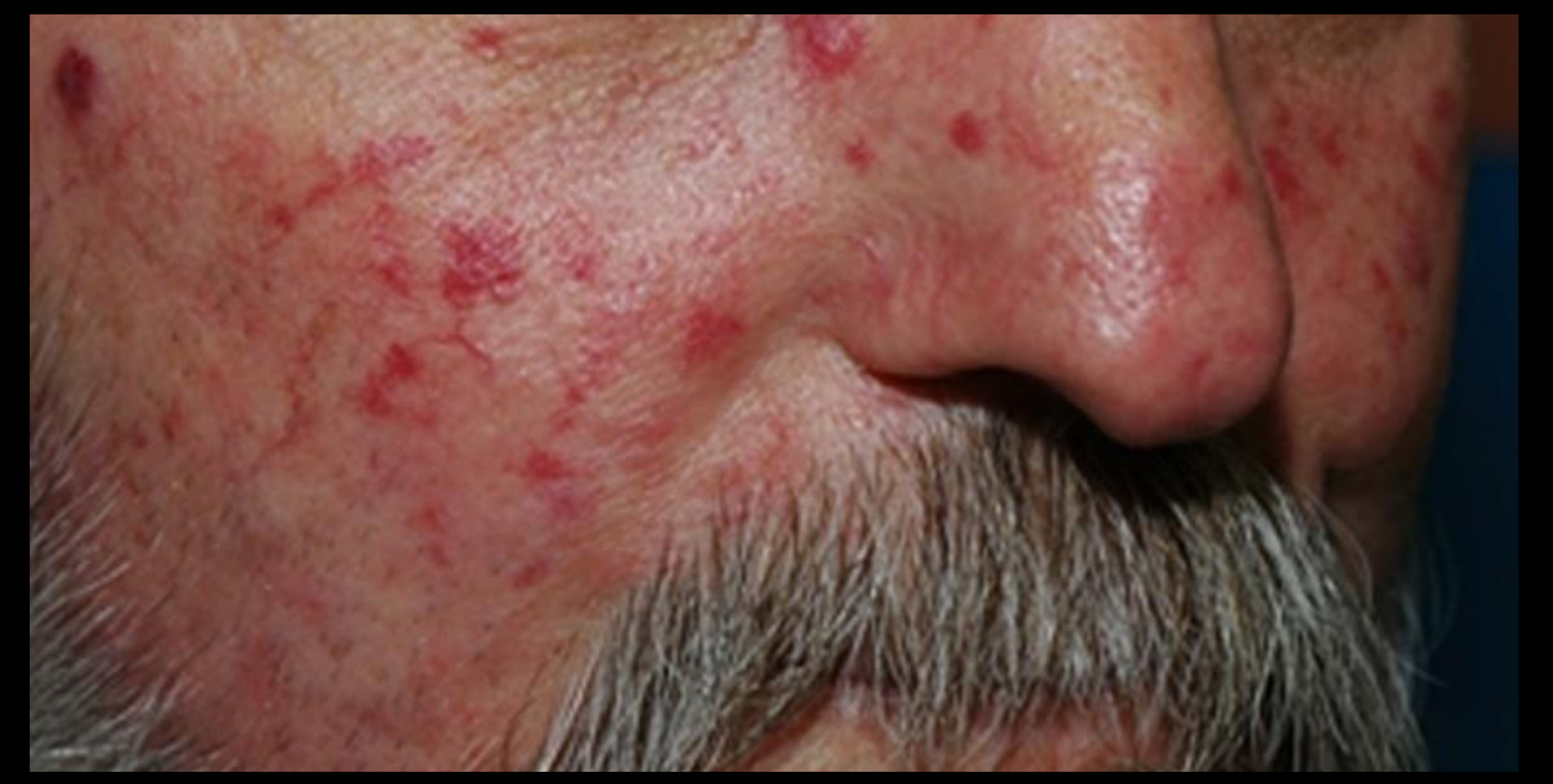




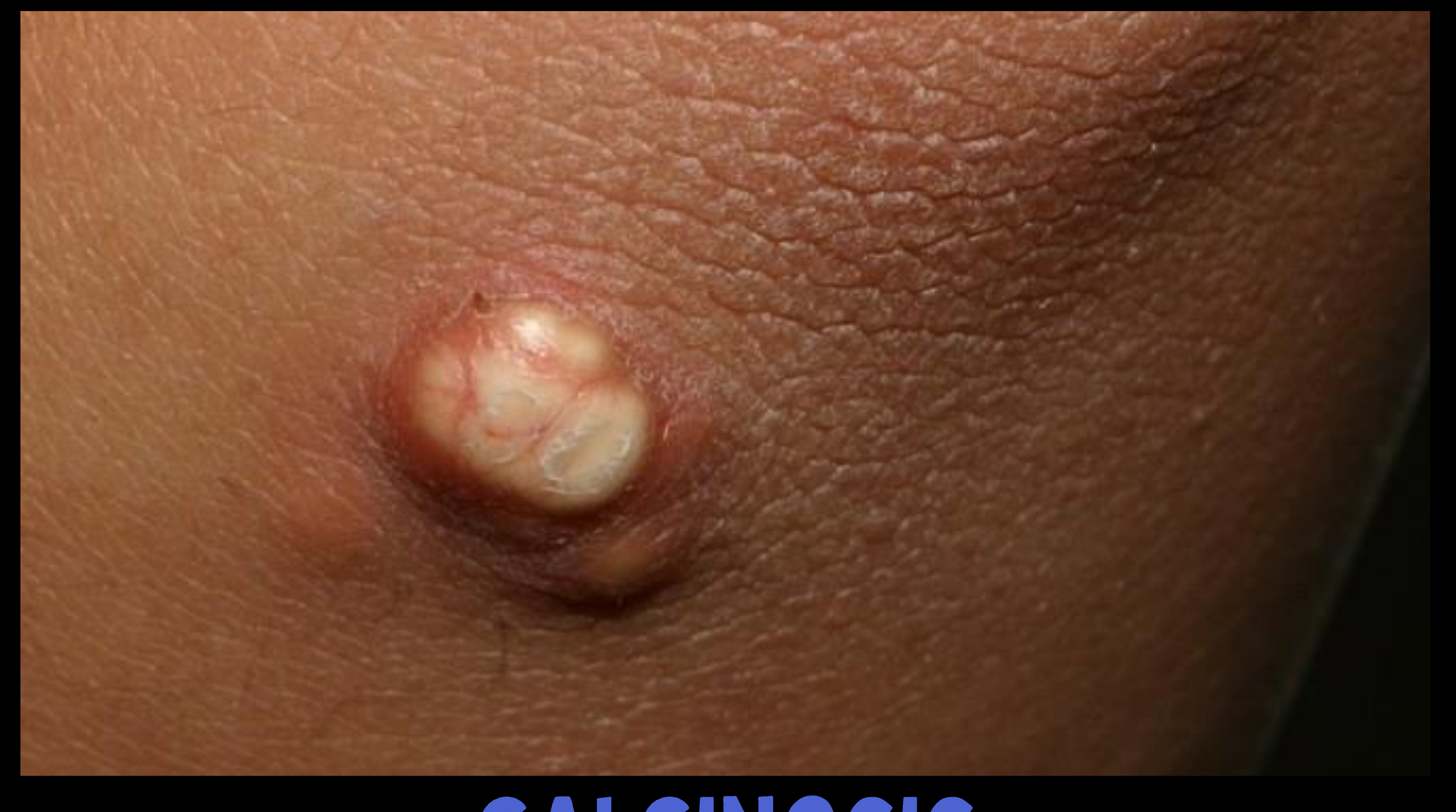
SALT AND PEPPER SKIN

OTHER SKIN FINDINGS

PITS AND ULCERS

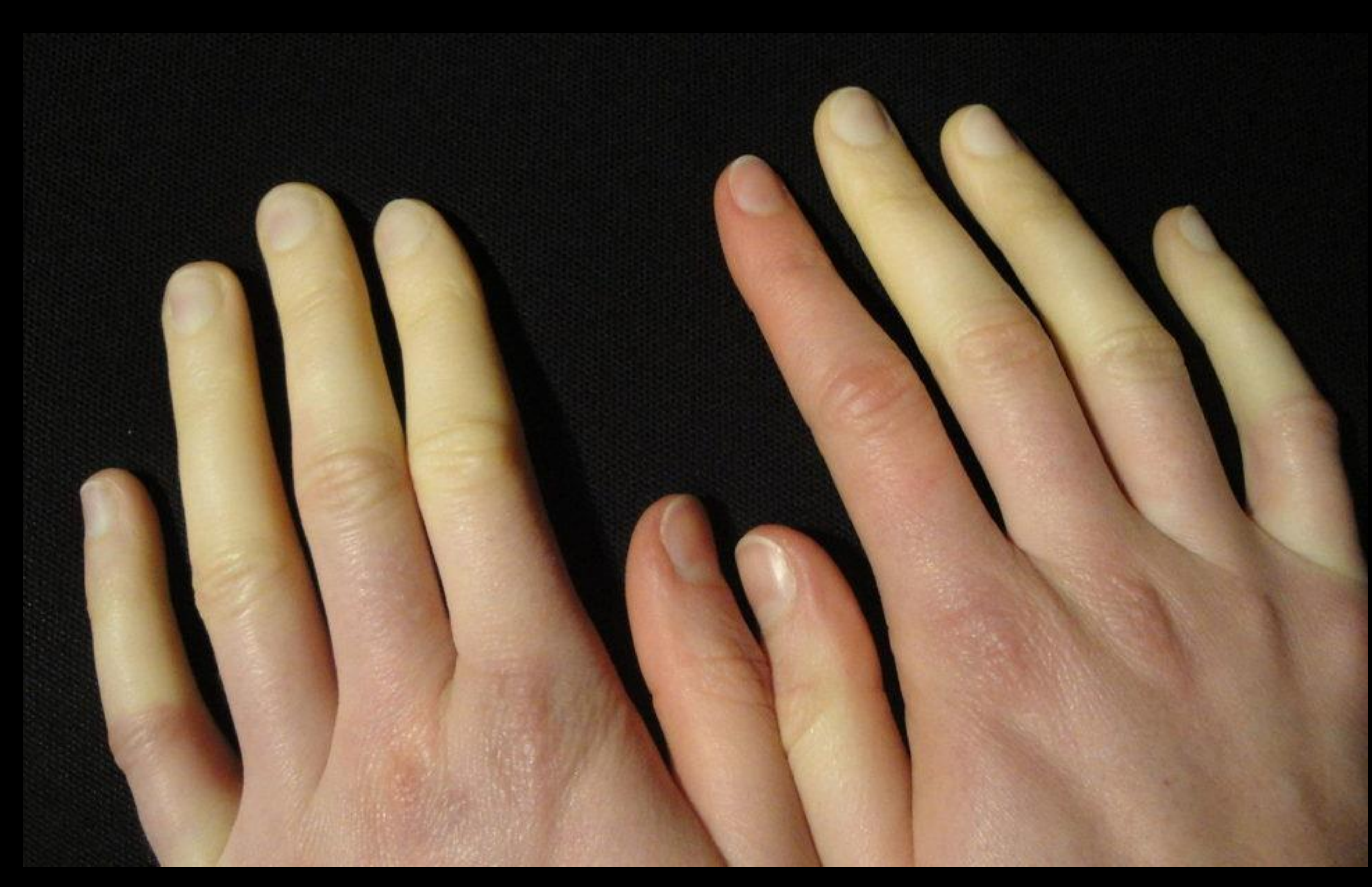


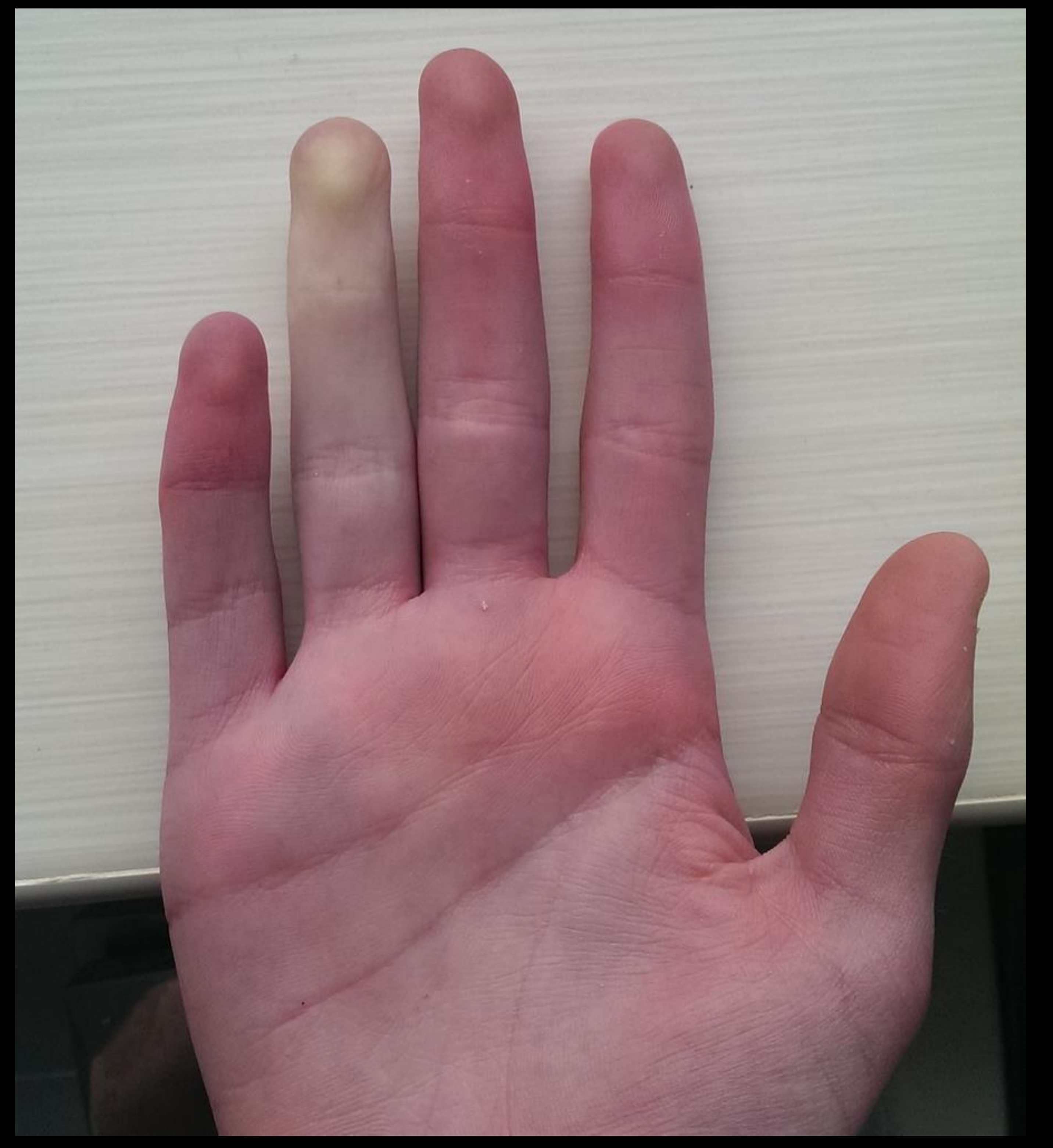
TELANGIECTASIAS



CALCINOSIS

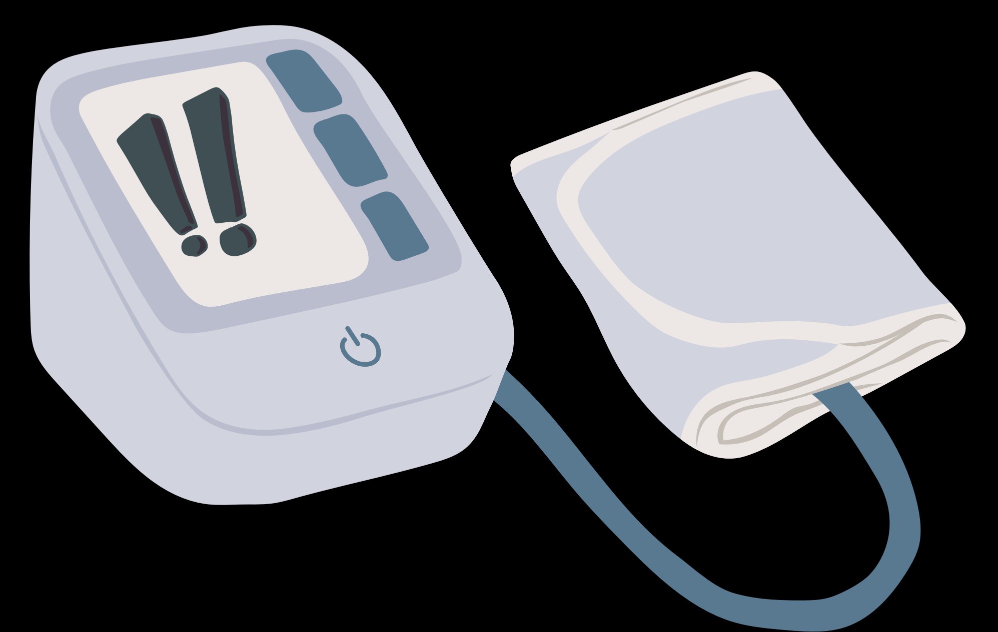
RAYNAUD'S 95%



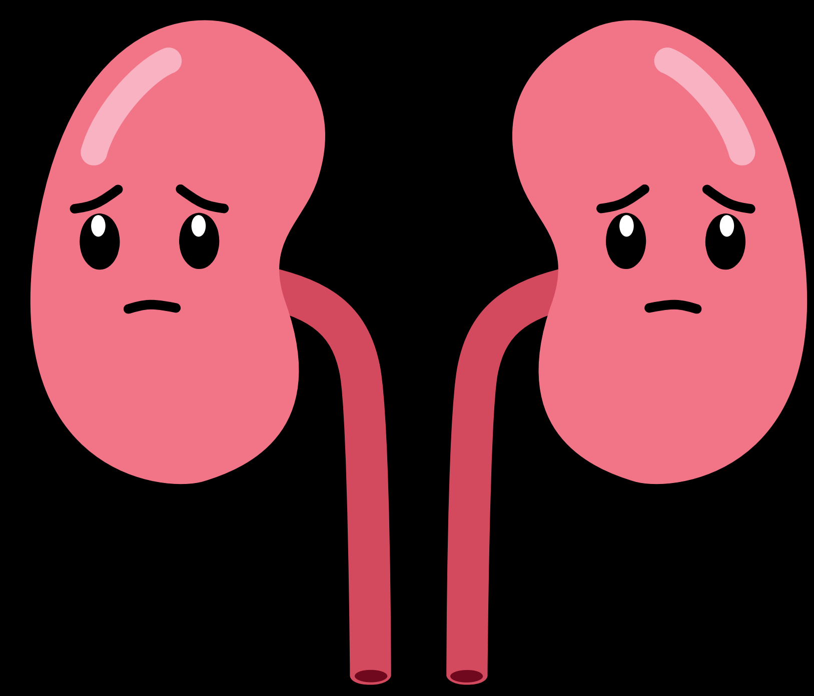


SCLERODERMA RENAL CRISIS

10% OF PATIENTS WITH DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS



ACUTE ONSET OF SEVERE HYPERTENSION



RENAL INSUFFICIENCY

MICROANGIOPATHIC HEMOLYTIC ANEMIA

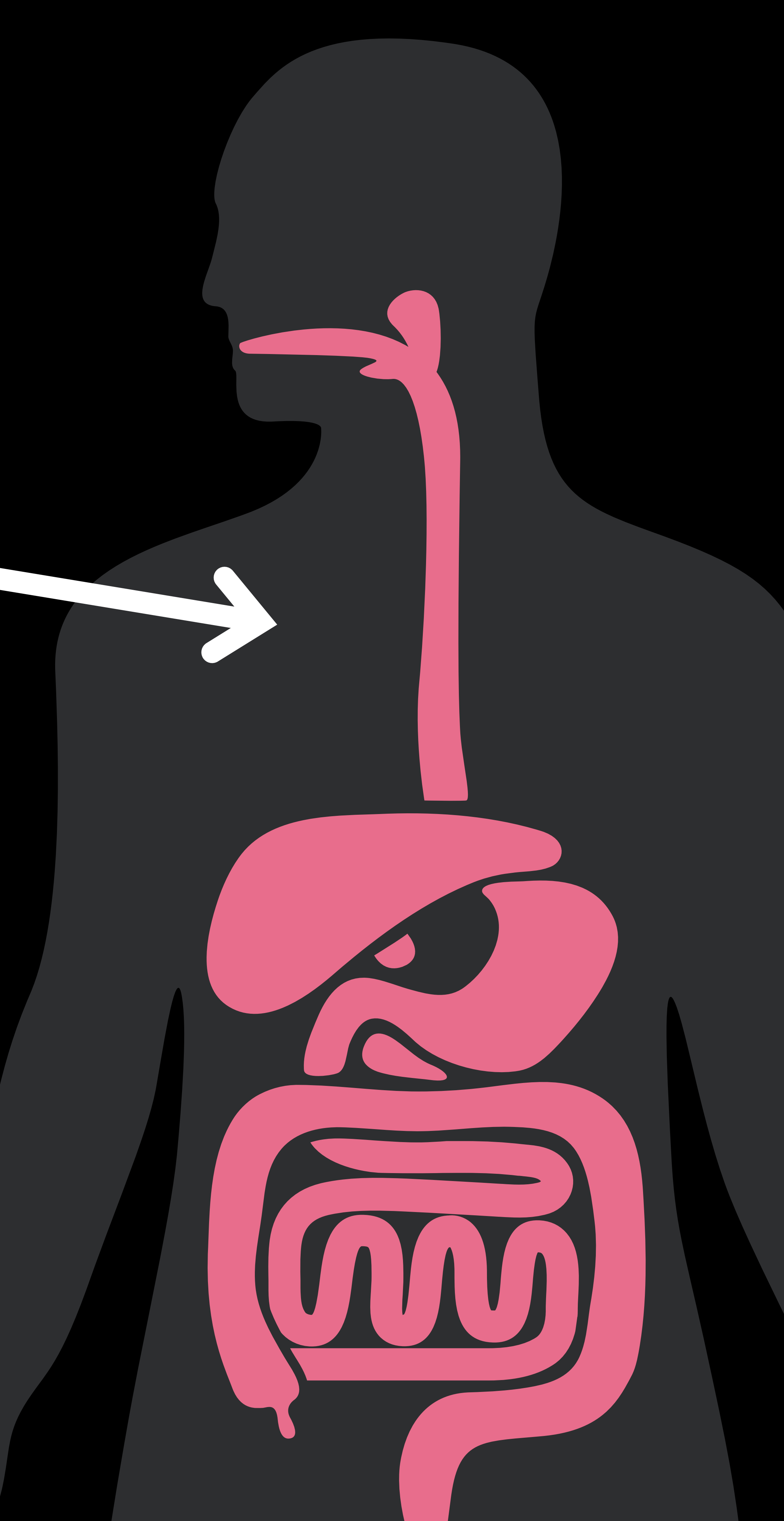
TREATMENT: ACE INHIBITORS

GASTROINTESTINAL

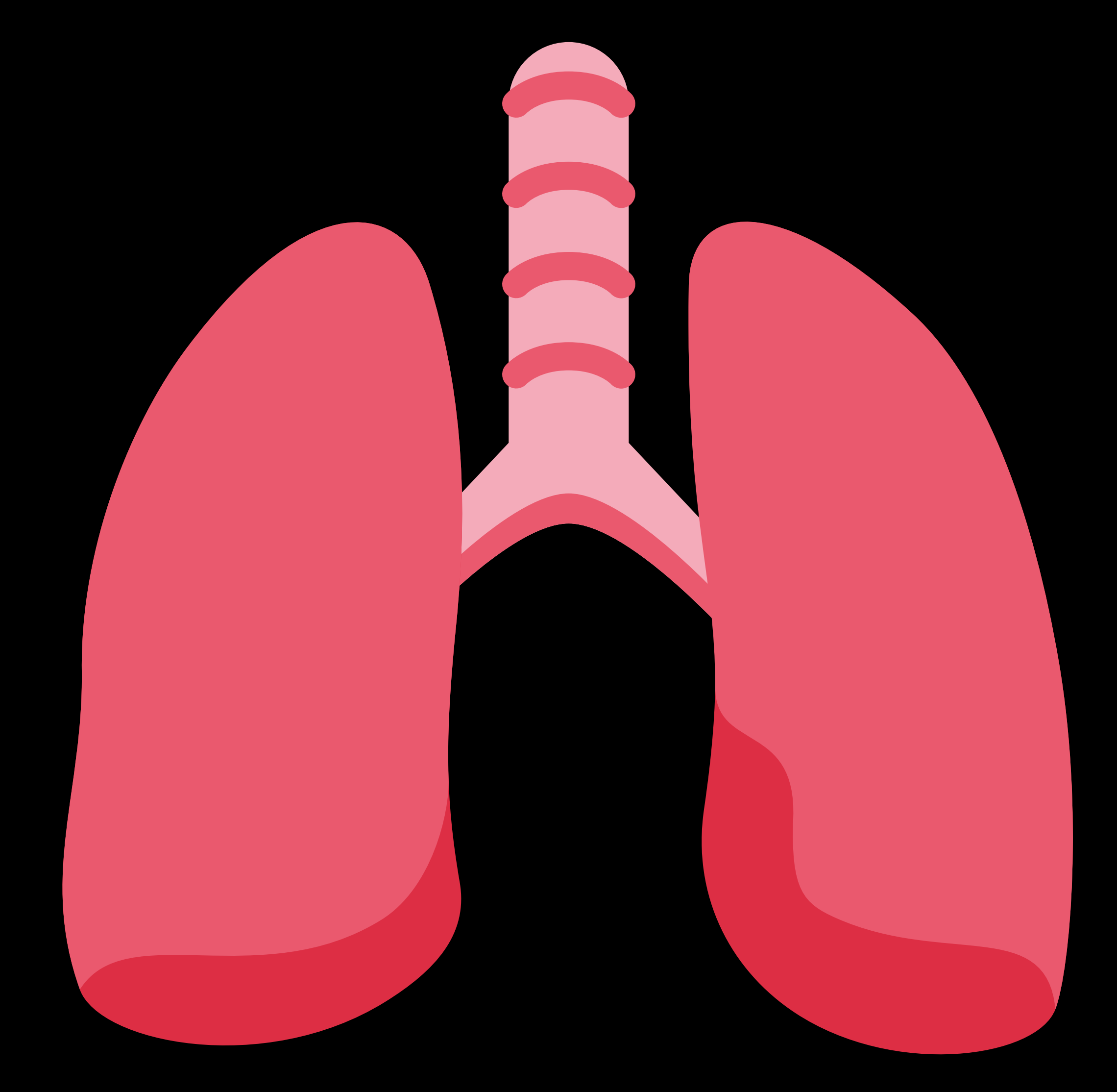
80% ESOPHAGUS

DYSPHAGIA

- GERD
- ESOPHAGITIS
- STRICTURE
- BARRETT'S ESOPHAGUS
- GASTRIC TELANGIECTASIAS
- GI BLEEDING
- ILEUS



PULMONARY

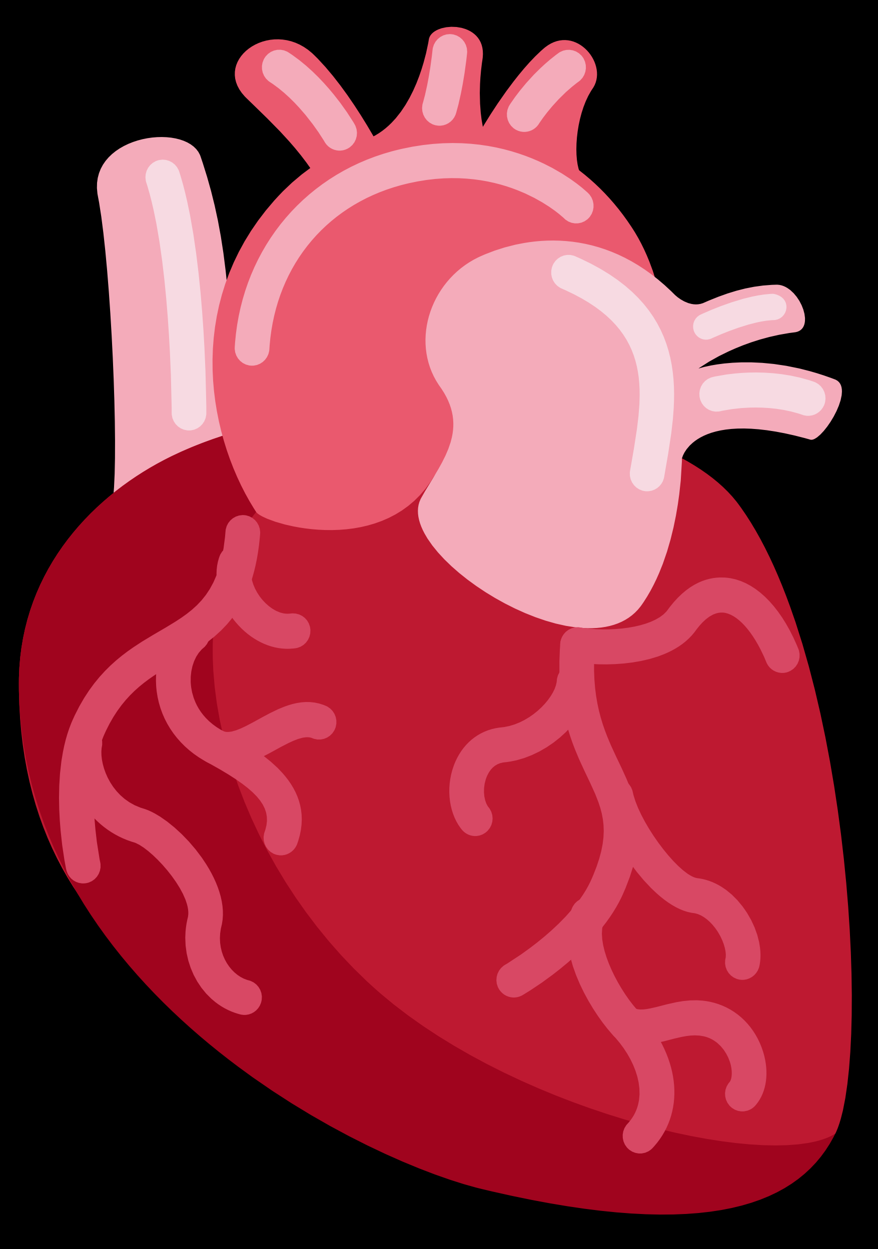


INTERSTITIAL LUNG DISEASE

PULMONARY FIBROSIS

PULMONARY ARTERIAL HYPERTENSION

RIGHT HEART FAILURE

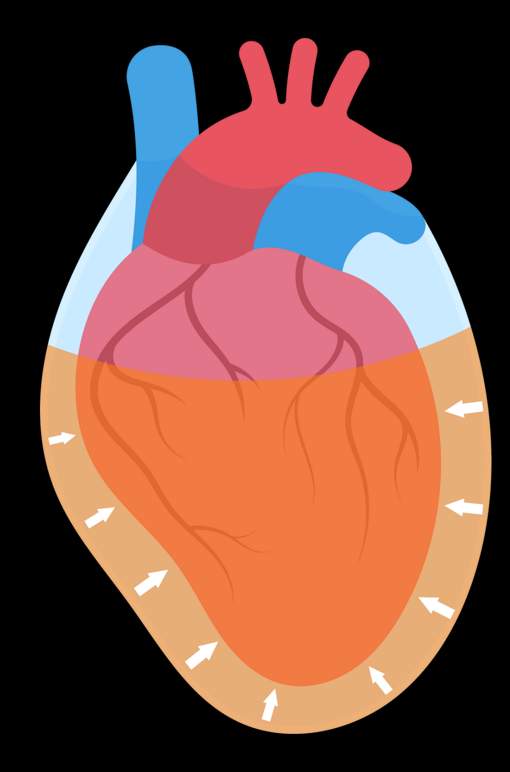


CARDIAC INVOLVEMENT

- PERCARDITIS
- PERICARDIAL EFFUSION
- DILATED CARDIOMYOPATHY
- DYSRHYTHMIA

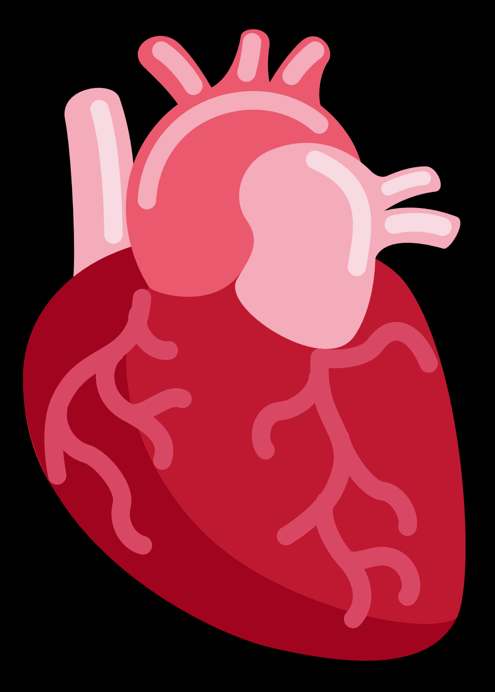
CARDIAC INVOLVEMENT

PERICARDIAL EFFUSION



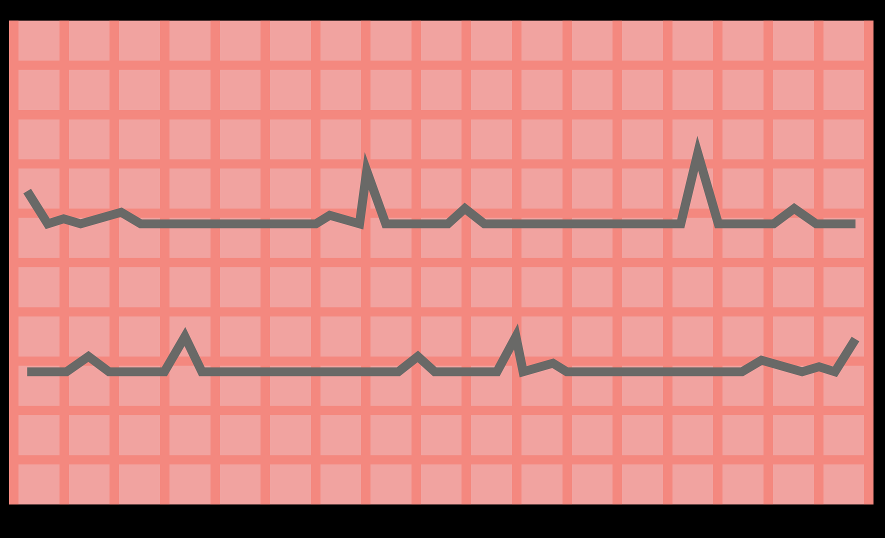
CAN HERALD ONSET OF SCLERODERMA RENAL CRISIS

DILATED CARDIOMYOPATHY



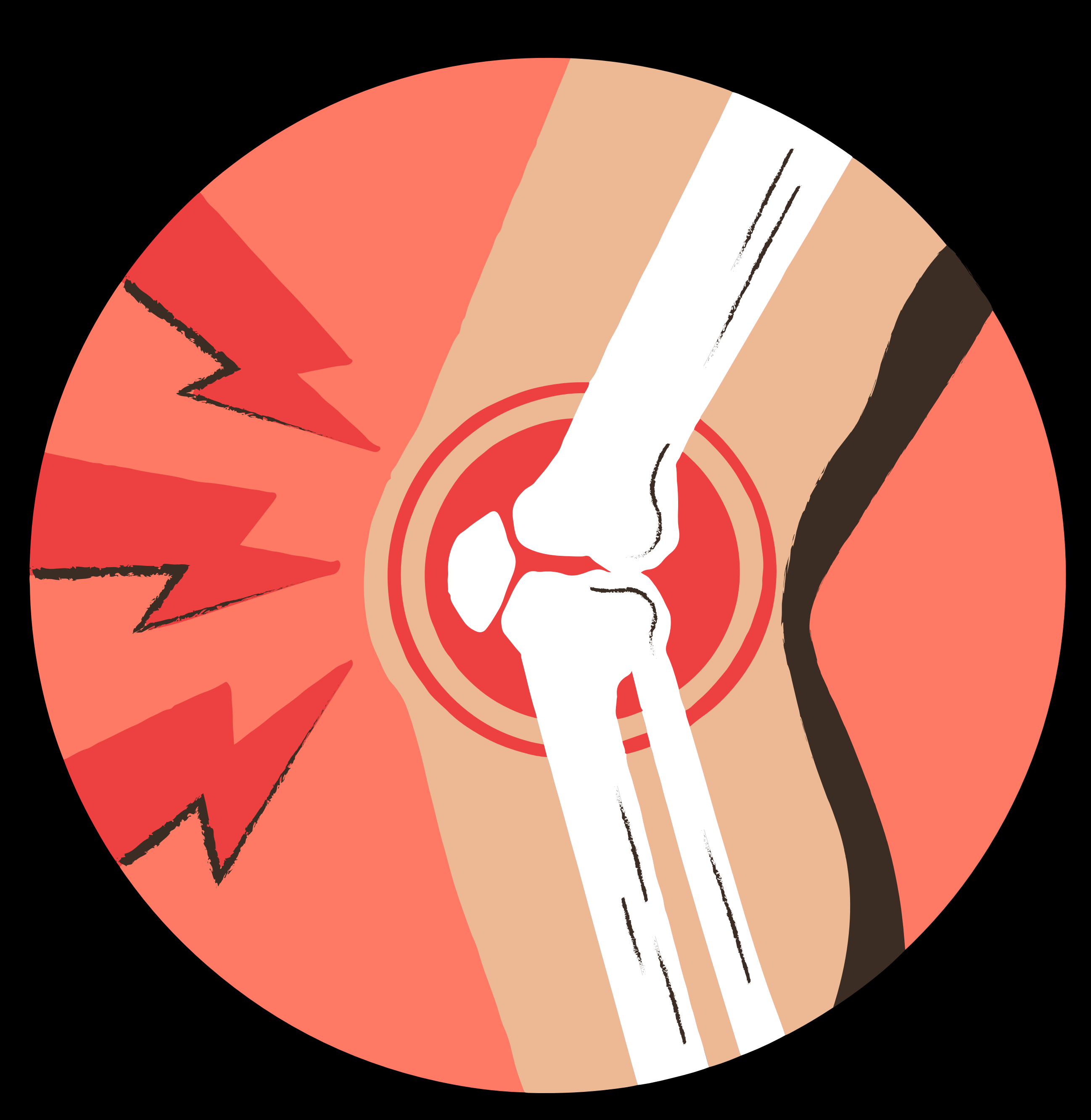
USUALLY RESULTS FROM PATCHY MYOCARDIAL FIBROSIS

DYSRHYTHMIA



FIBROSIS IN THE CONDUCTION PATHWAYS

MUSCULOSKELETAL



INFLAMMATORY ARTHRITIS

MYALGIA

MYOPATHY
COLLAGEN DEPOSITS

SYSTEMIC SCLEROSIS HAS THE HIGHEST MORTALITY AMONG AUTOIMMUNE DISEASES

"POOR ACCESS TO SPECIALTY CARE IS A MAJOR FACTOR DRIVING POOR OUTCOMES IN SYSTEMIC LUPUS ERYTHEMATOSUS (SLE). SLE PATIENTS WHO ARE RACIAL/ETHNIC MINORITIES, HAVE LOW SOCIOECONOMIC STATUS, AND WITH PUBLIC INSURANCE FACE DIFFICULTIES IN ACCESSING SPECIALTY CARE."

ACCESS TO CARE

USE OF QUALITY MEASURES TO IDENTIFY DISPARITIES IN HEALTH CARE FOR SYSTEMIC LUPUS ERYTHEMATOSIS

REACTIVE ARTHRITIS

CHALLENGE: DIAGNOSIS

REACTIVE ARTHRITIS

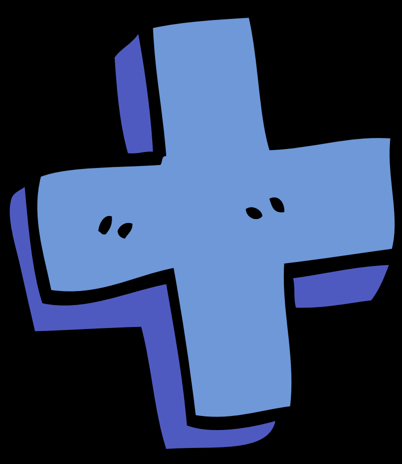
FORMERLY REITER'S SYNDROME

INFLAMMATORY
ARTHRITIS

DAYS TO WEEKS
AFTER GI OR GU
INFECTION

EYE
MUCOSA
OFTEN INVOLVED

GENETIC PREDISPOSITION



TRIGGER

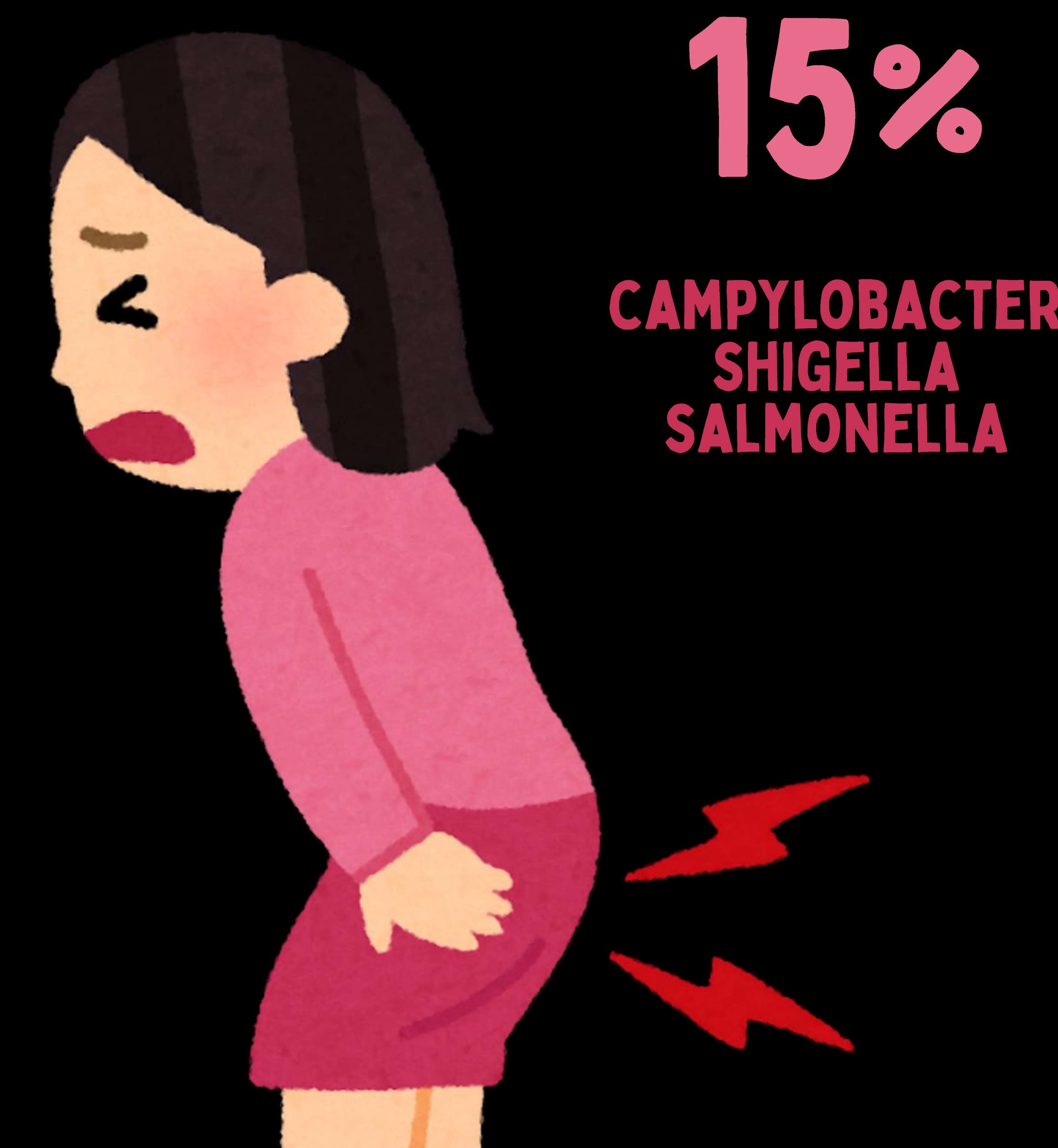
INFECTION OR OTHER
ENVIRONMENTAL TRIGGER

GENETIC PREDISPOSITION
HLA B27

2%

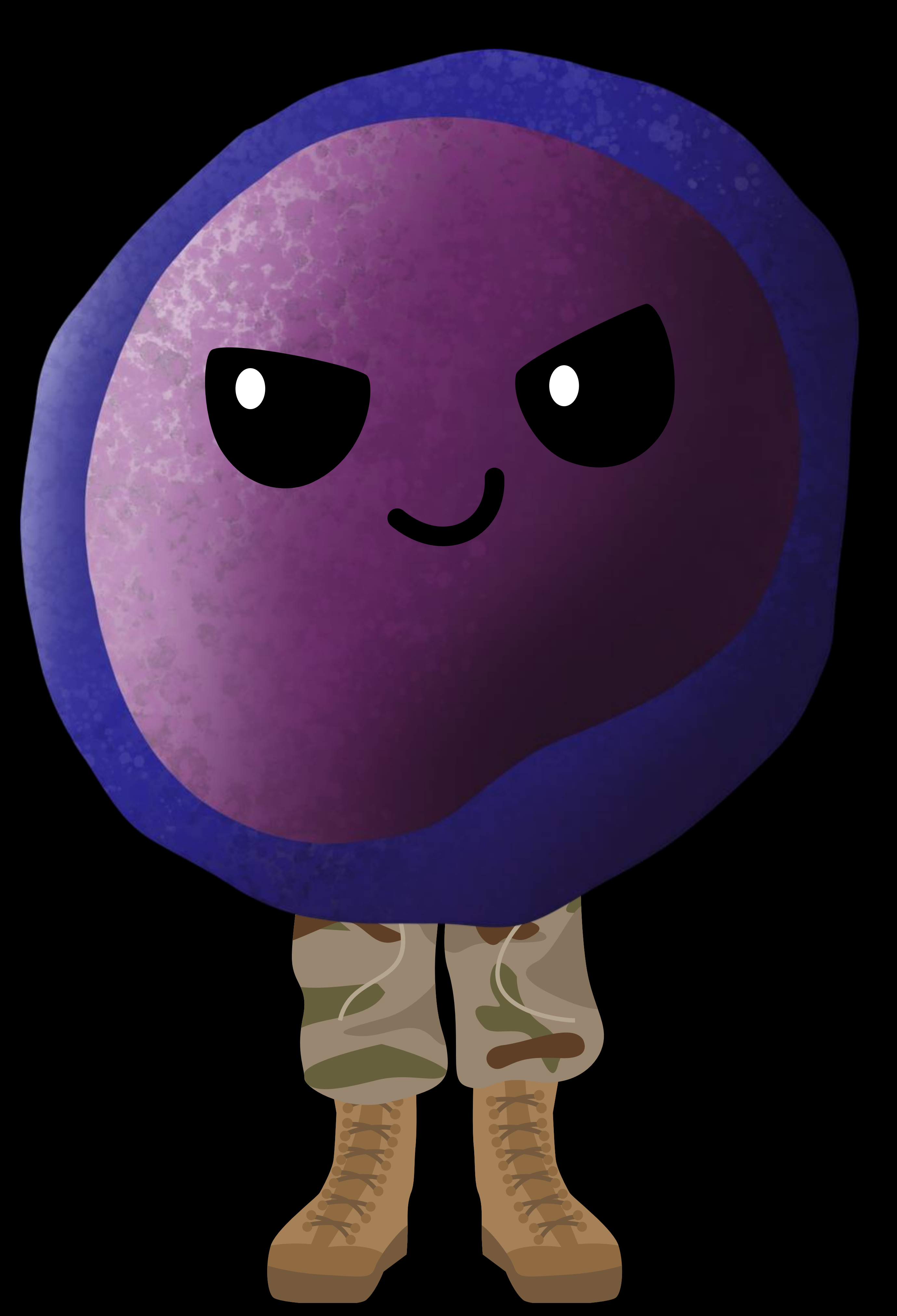
AFTER CHLAMYDIA INFECTION

AFTER GASTROENTERITIS

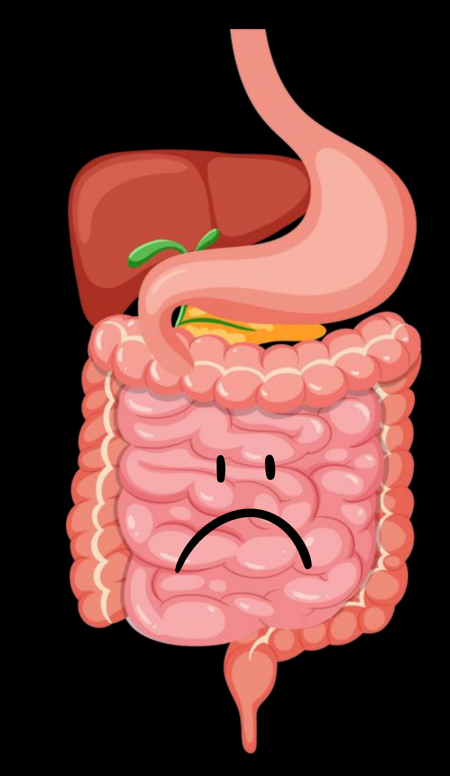


15%

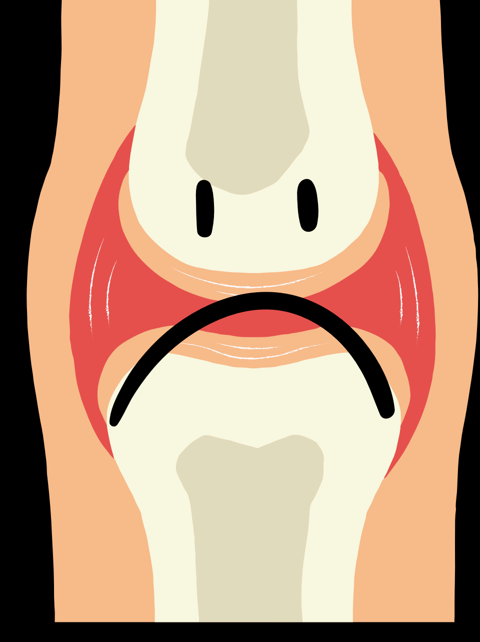
CAMPYLOBACTER
SHIGELLA
SALMONELLA



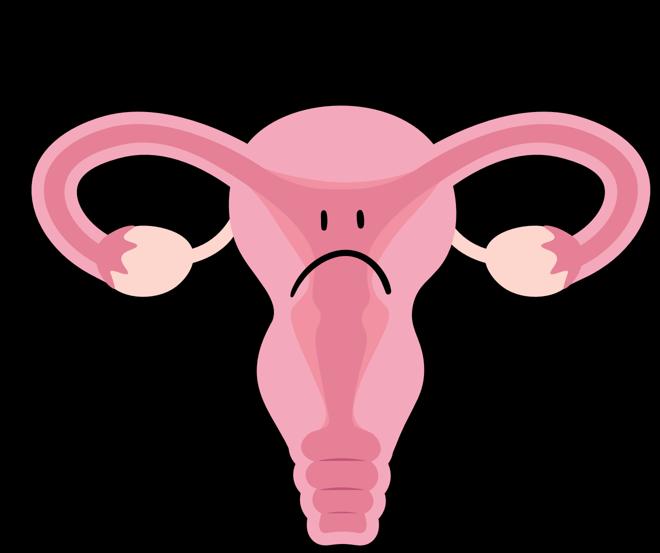
AUTOANTIBODIES
ATTACK THE
SYNOVIUM
AND OFTEN OTHER
TISSUES



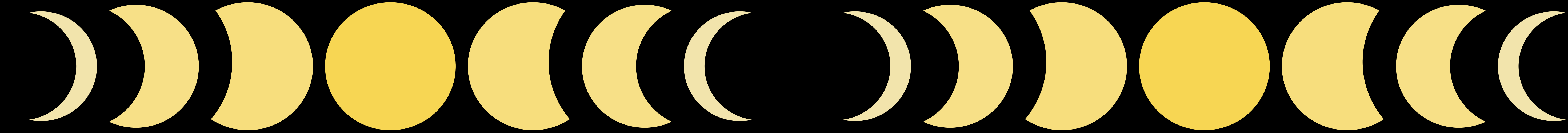
BACTERIAL
GASTROENTERITIS



INFLAMMATORY
ARTHRITIS

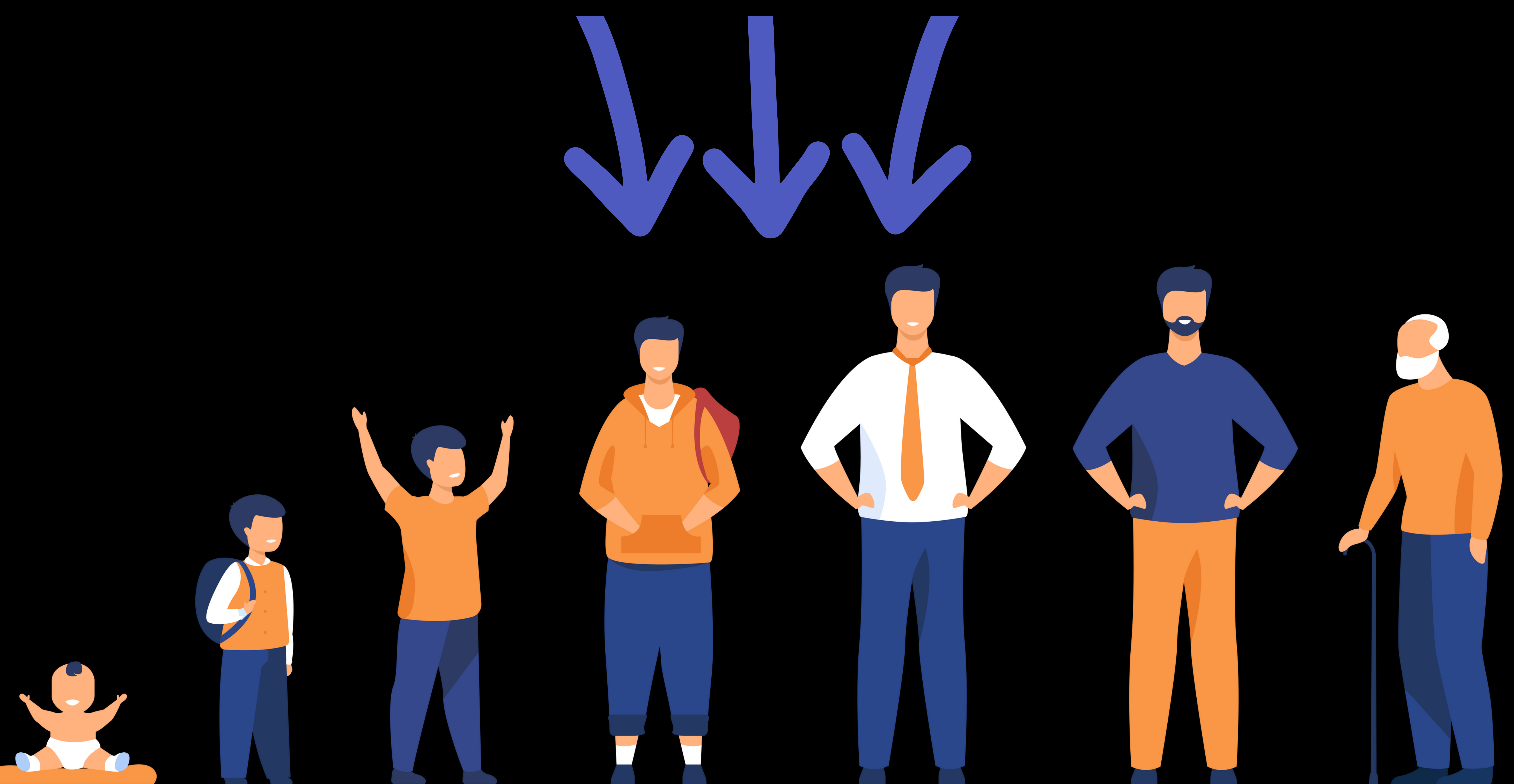


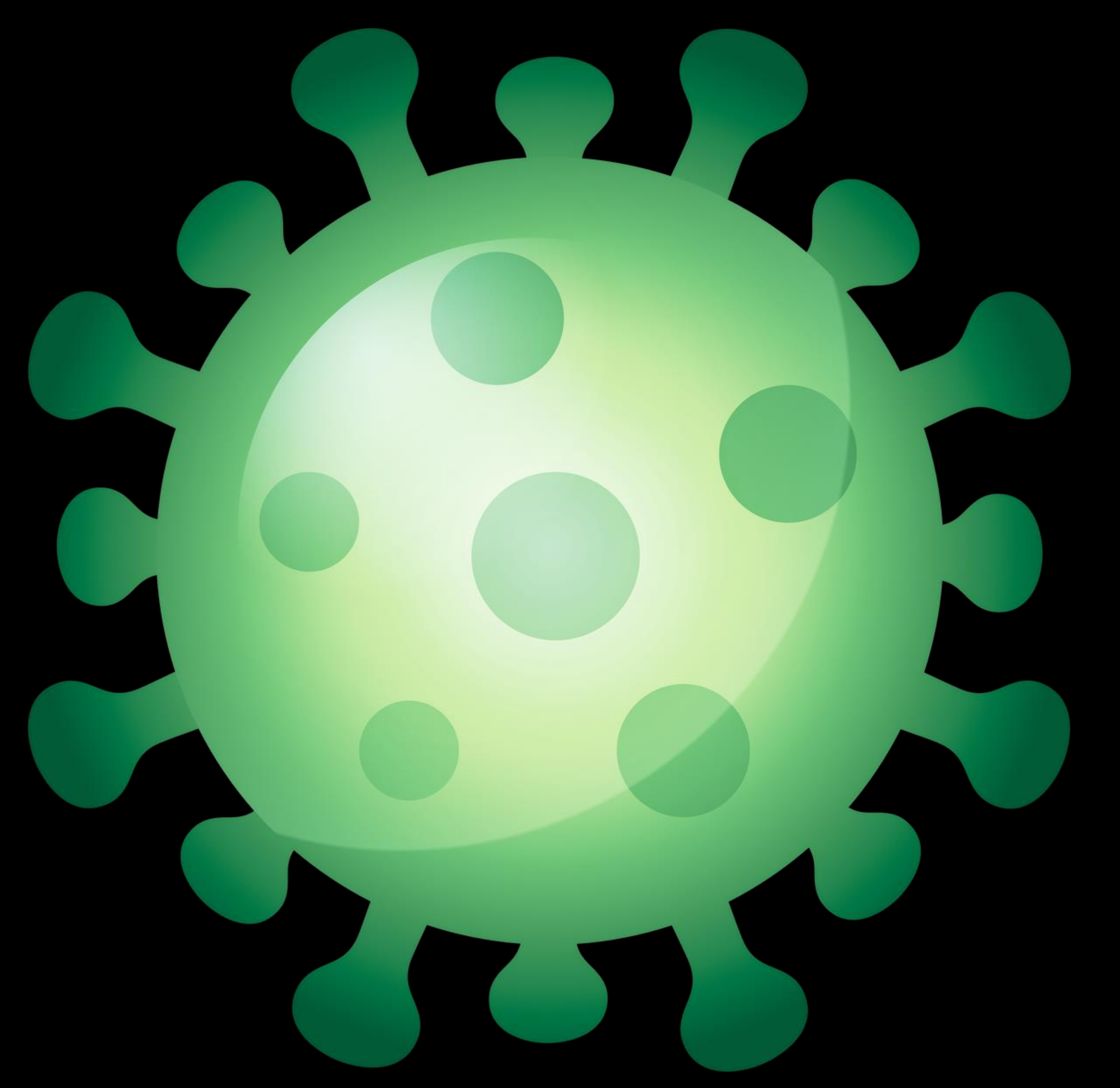
NON-GONOCOCCAL
CERVICITIS/URETHRITIS



ONSET WITHIN 2 MONTHS

MEN IN 2ND AND 3RD DECADES OF LIFE

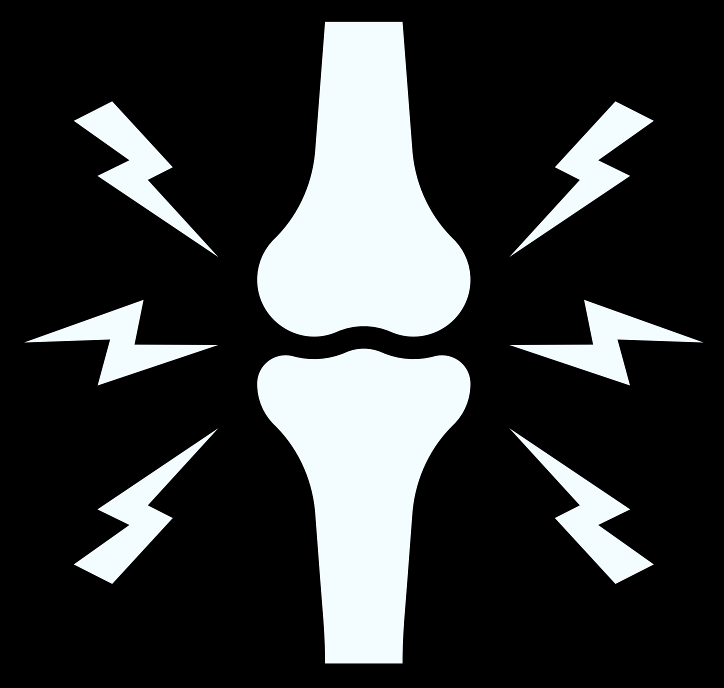




MUCH MORE COMMON IN PATIENTS WITH

HIV

YOUR PATIENT WILL PRESENT WITH



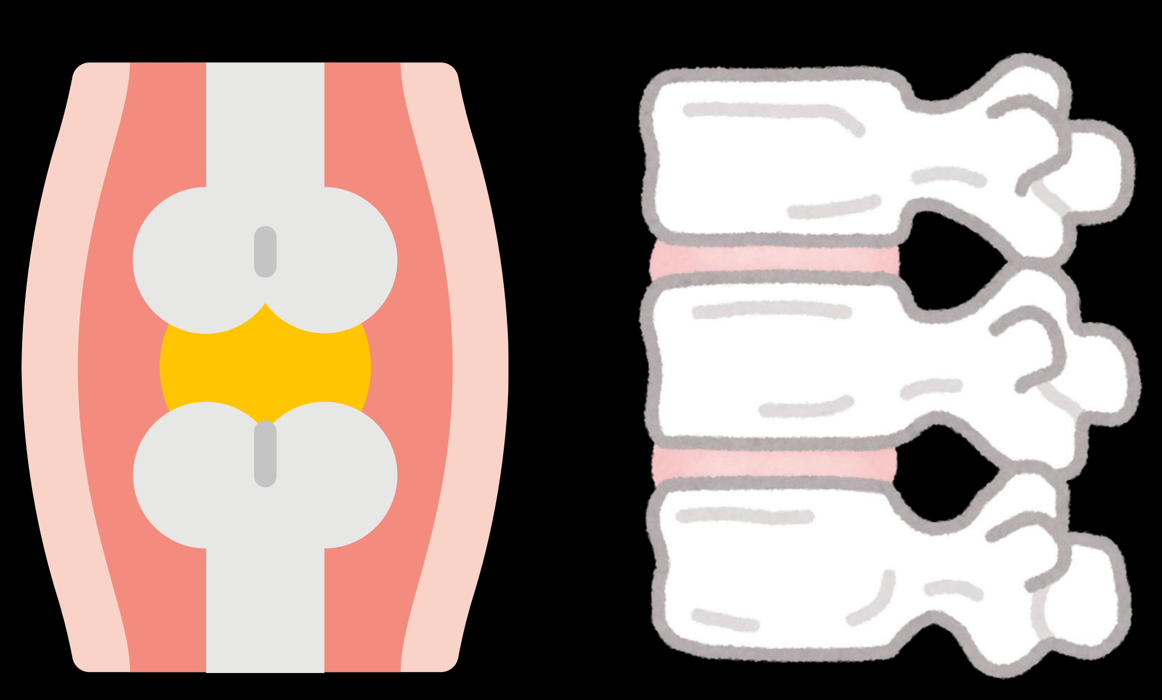
PAIN IN LARGE JOINTS

ASYMMETRIC

WORSE PAIN AT NIGHT

MORNING STIFFNESS

LOW BACK PAIN
+
LOWER EXTREMITY JOINT PAIN

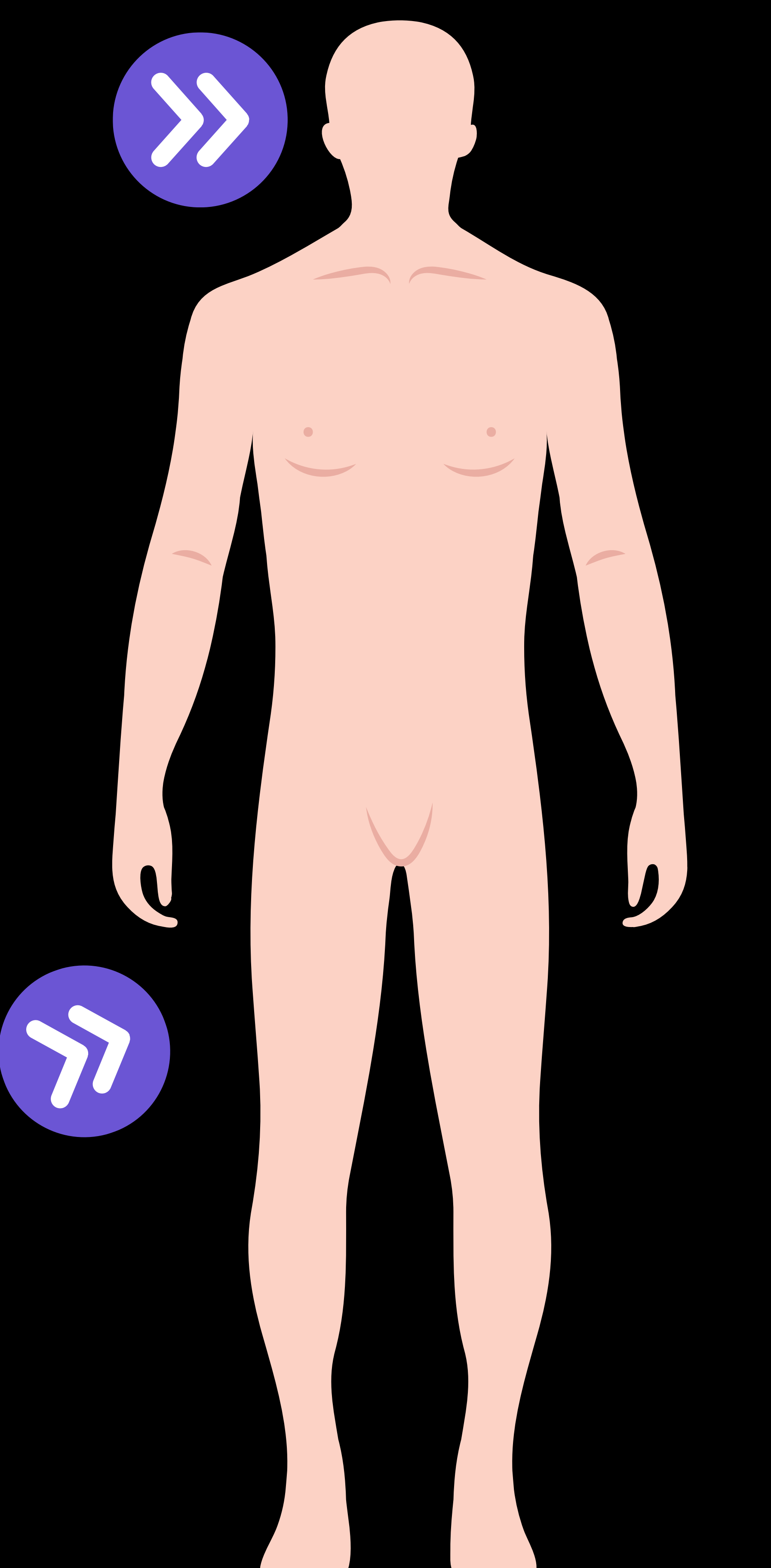


LUMBAR SPINE

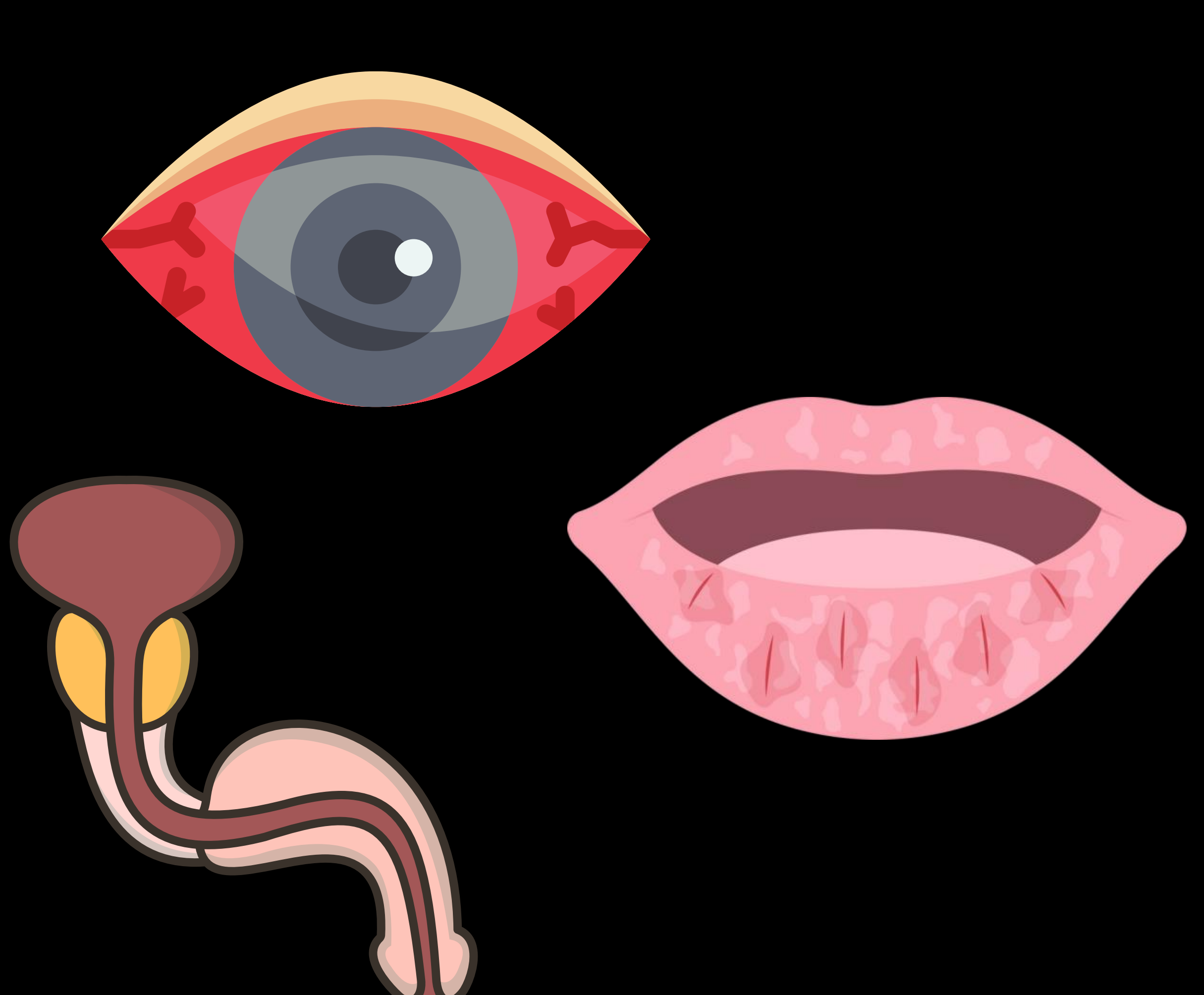
SI JOINT

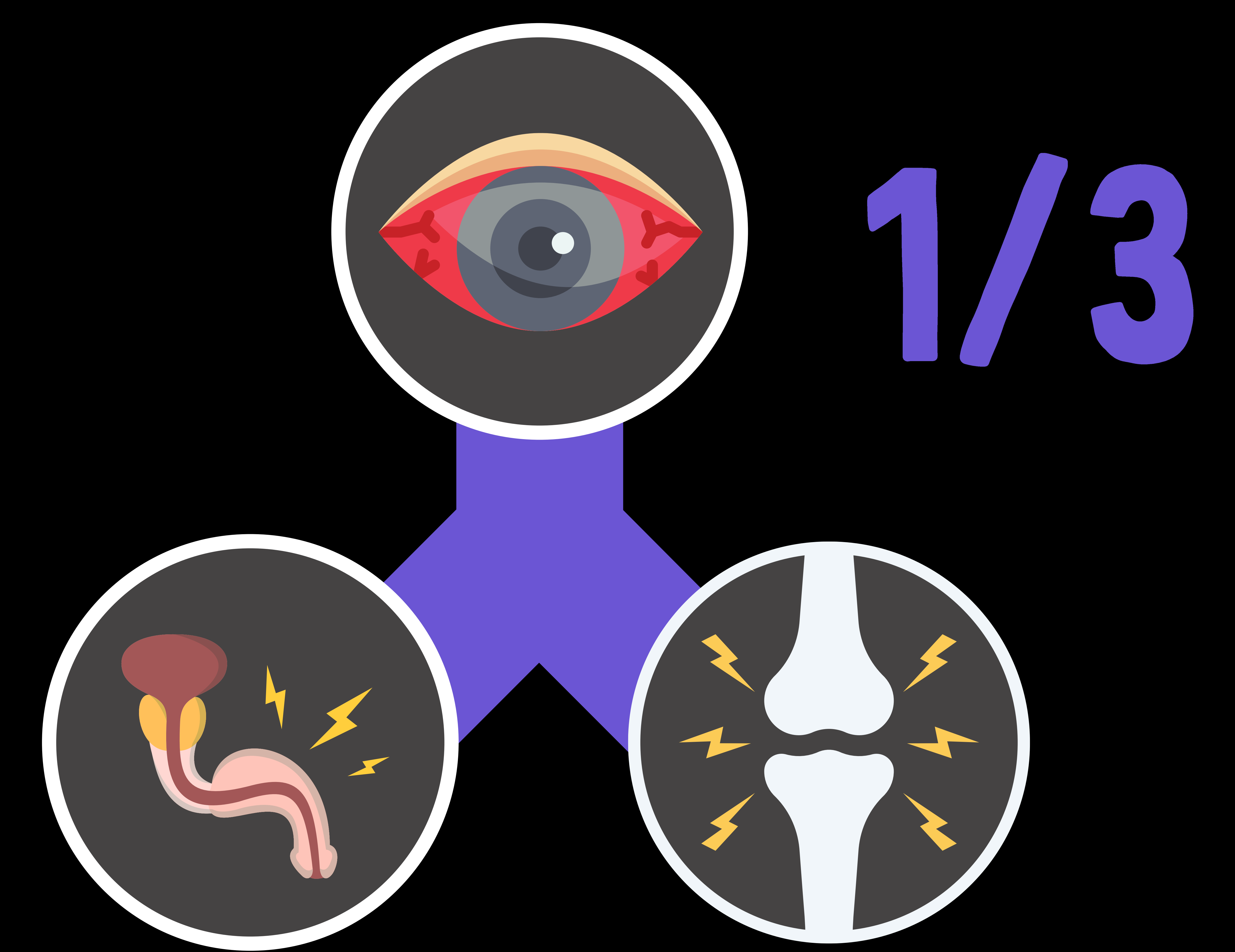
HEEL PAIN

CONJUNCTIVITIS
UVEITIS
MUCOCUTANEOUS LESIONS



MAY PRECEDE ARTHRITIS




1/3

2/3

DO NOT!

1/3

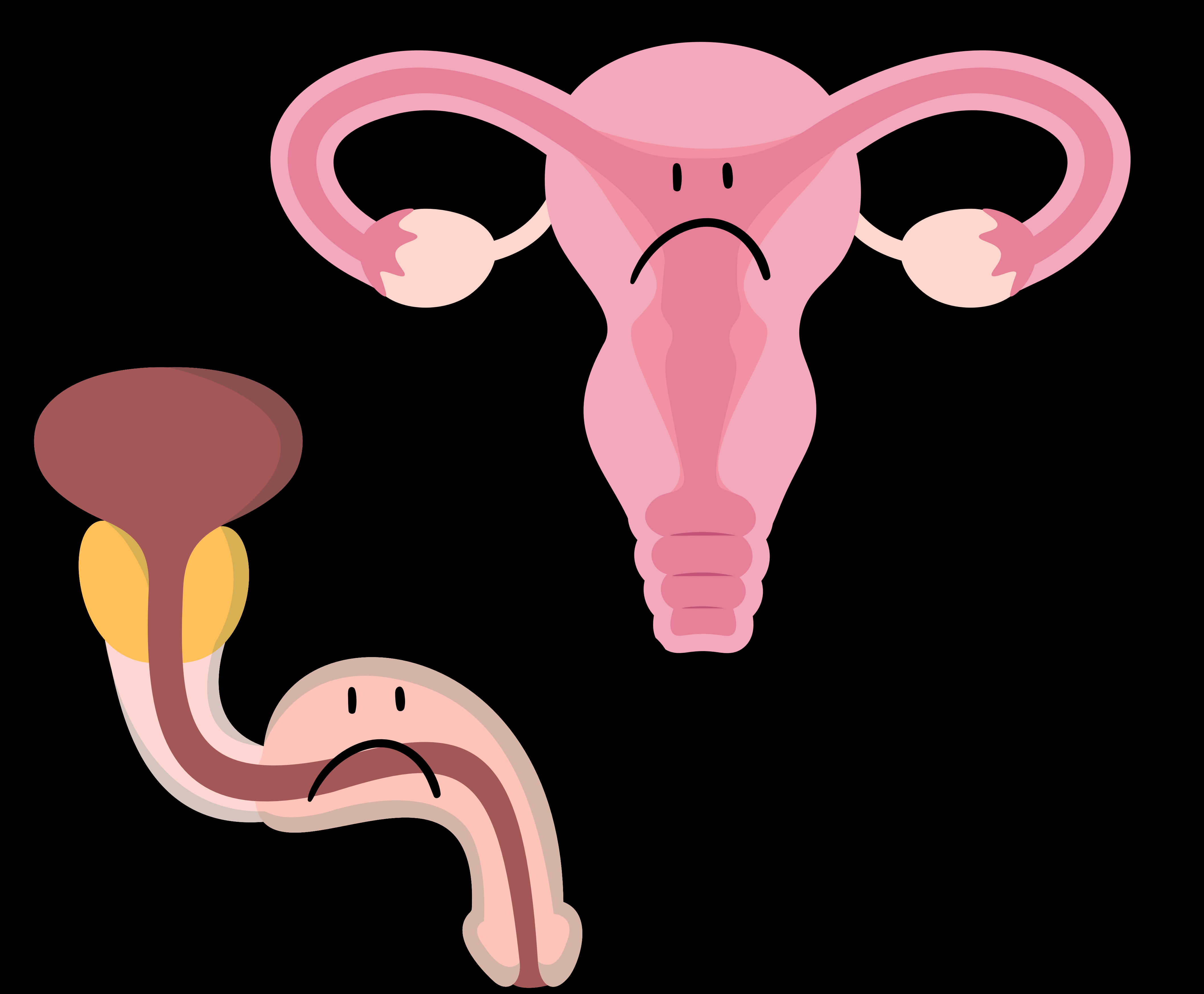
OTHER FINDINGS

- ENTHESITIS
- DACTYLITIS
- EPISCLERITIS
- KERATITIS
- PROSTATITIS
- SALPINGO-OOPHORITIS
- CYSTITIS
- BALANITIS
- MOUTH ULCERS
- ERYTHEMA NODOSUM
- CARDITIS
- AORTIC VALVE REGURG
- DYSRHYTHMIA
- NAIL PITS
- ONYCHOLYSIS
- NAIL DYSTROPHY
- PUSTULAR PSORIASIS
- GEOGRAPHIC TONGUE

REACTIVE ARTHRITIS

YOU'VE GOT TO REMEMBER THIS IN YOUR DIFFERENTIAL

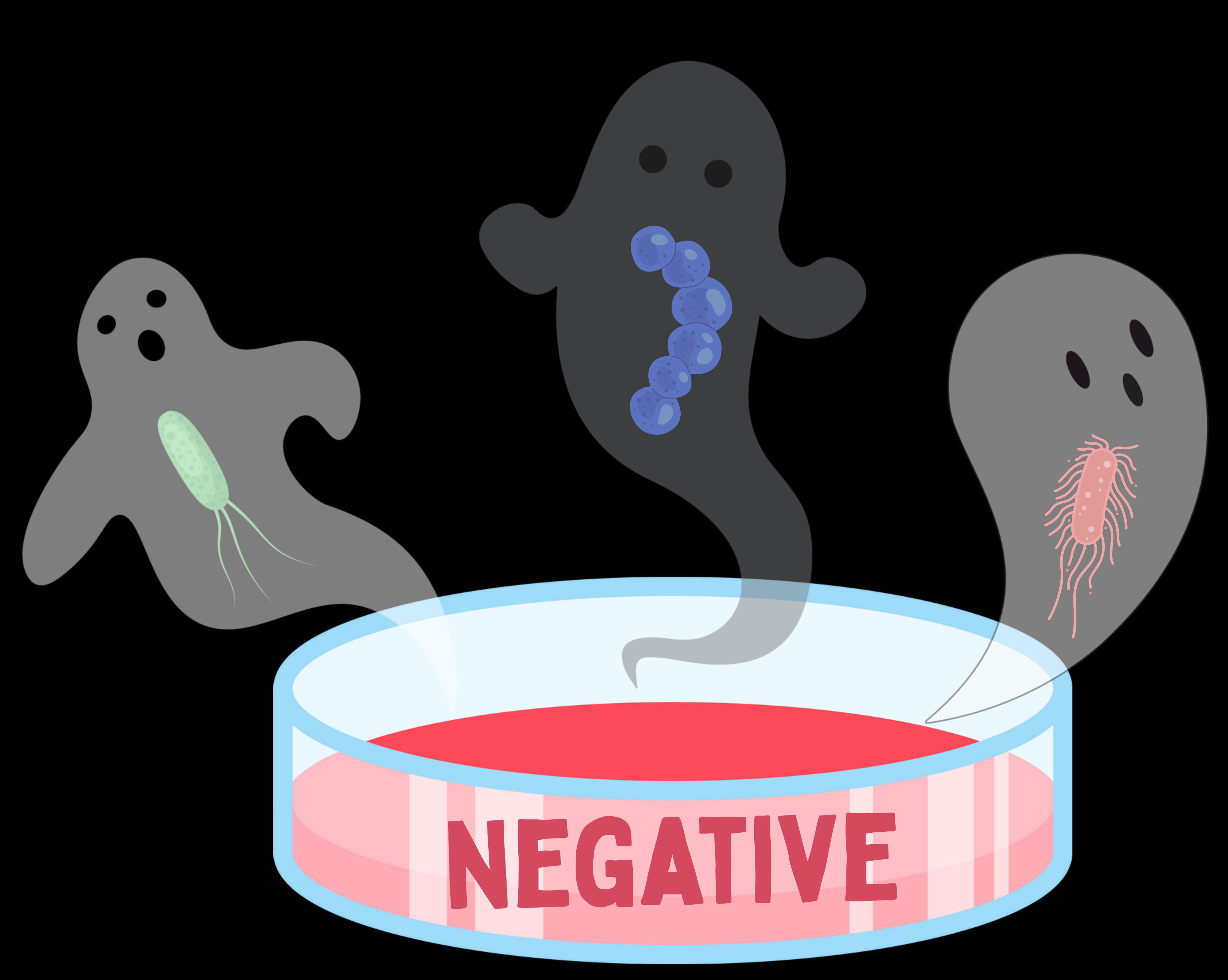
YOU'VE GOT TO ASK THE RIGHT QUESTIONS



CHLAMYDIA CERVICITIS AND URETHRITIS ARE OFTEN ASYMPTOMATIC

BY THE TIME ARTHRITIS DEVELOPS

CULTURES FOR THE BACTERIAL TRIGGER ARE OFTEN



HUNTIN' VAPORS

ESTABLISHING THE DIAGNOSIS

NEED 2

DACTYLITIS
ENTHESITIS
UVEITIS
OLIGOARTHRITIS
URETHRITIS OR GENITAL ULCERS
REPORTED DIARRHEA OR CERVICITIS WITHIN 4 WEEKS

PLUS

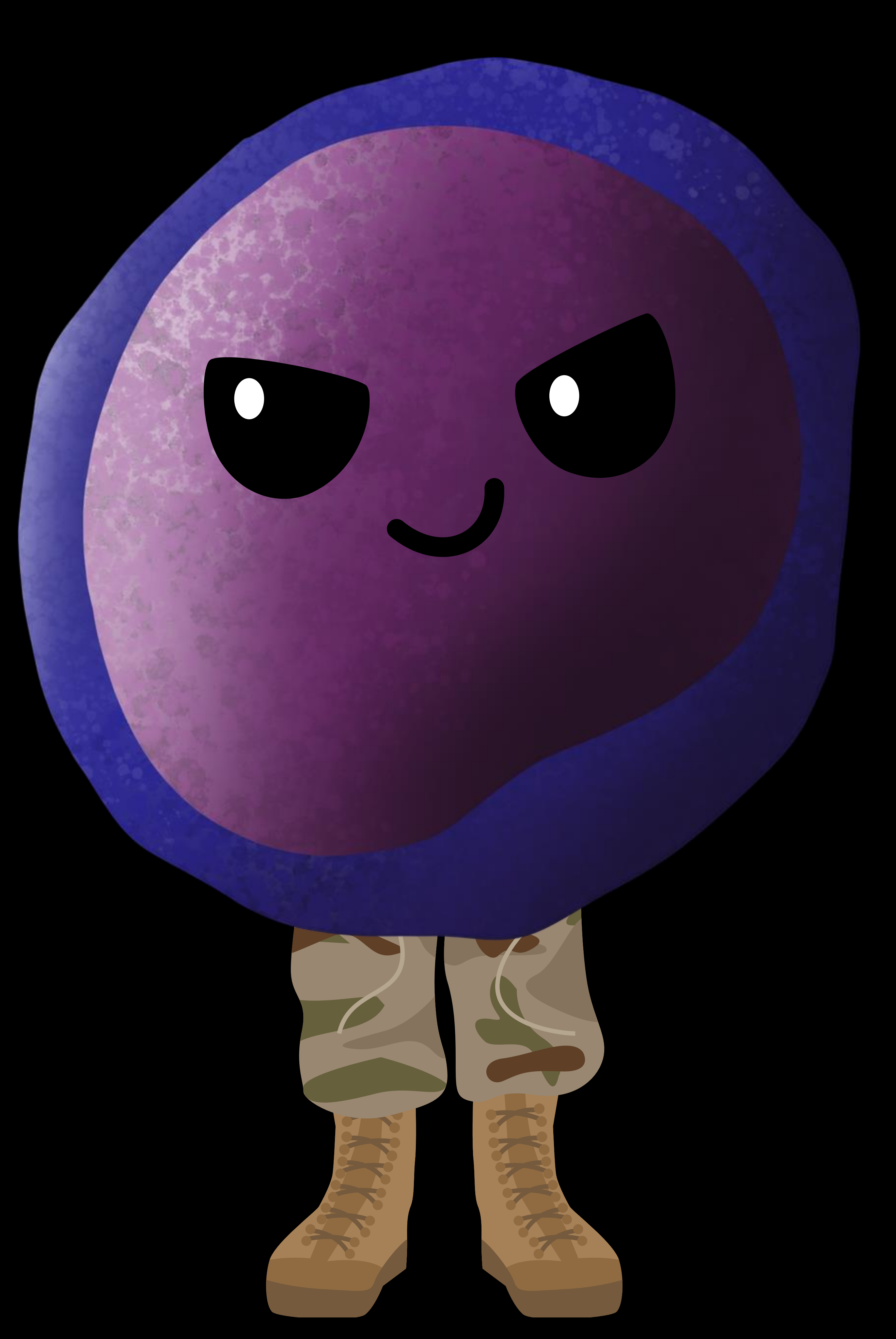
INVOLVEMENT OF SKELETAL SYSTEM

TREATMENT

- ANTIBIOTICS IF THE OFFENDING INFECTION IS STILL PRESENT
- NSAIDS
- PHYSICAL THERAPY
- DMARDS MAY BE HELPFUL

SYMPTOMS TYPICALLY LAST 4-6 MONTHS

UP TO 25% MAY PROGRESS TO CHRONIC DISEASE



TAKE HOME POINTS

LUPUS NEPHRITIS

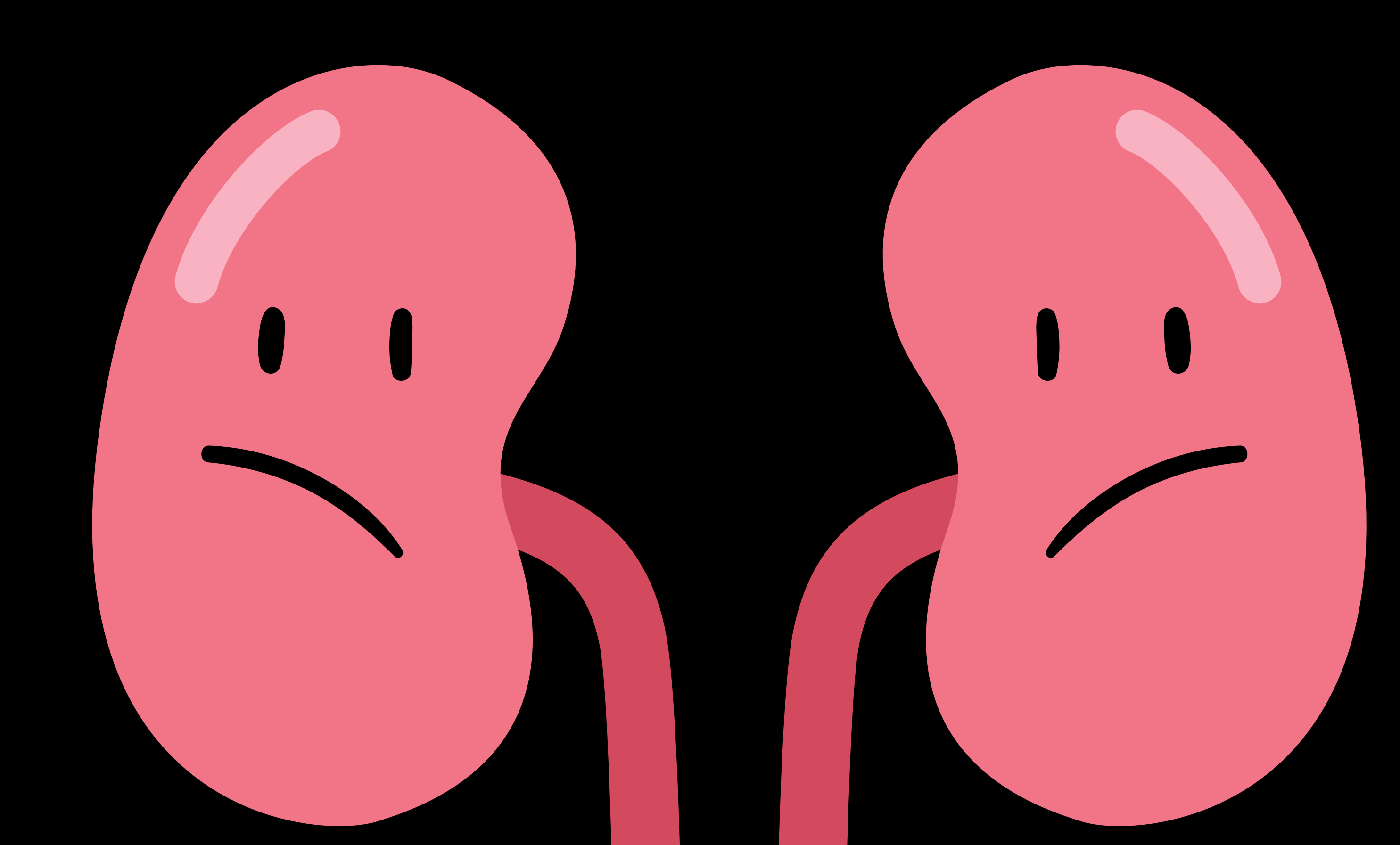
PRESENTS WITH HYPERTENSION AND EDEMA

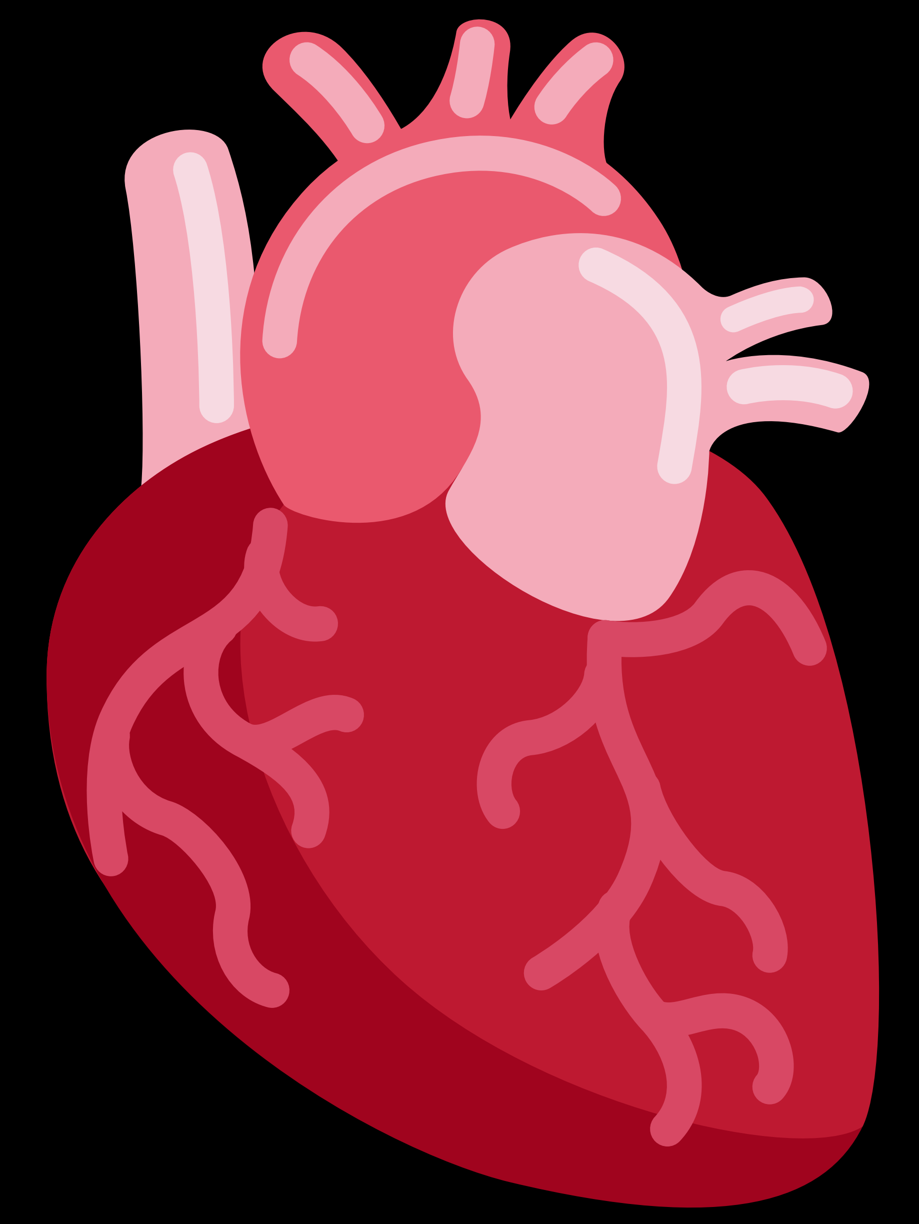
ABNORMAL URINALYSIS

RENAL FUNCTION MAY NOT BE ABNORMAL

CAN GO SOUTH QUICKLY

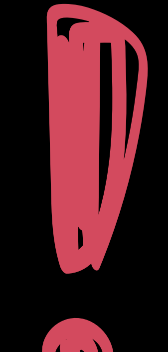
ADMIT FOR RENAL BIOPSY
AND STEROIDS





PATIENTS WITH SLE ARE AT **ESPECIALLY HIGH RISK** FOR CORONARY ARTERY DISEASE

YOUR 18- TO 39-YEAR-OLD LUPUS PATIENT HAS A HIGHER RISK OF HEART ATTACK AND STROKE THAN YOUR 50- TO 60-YEAR-OLD

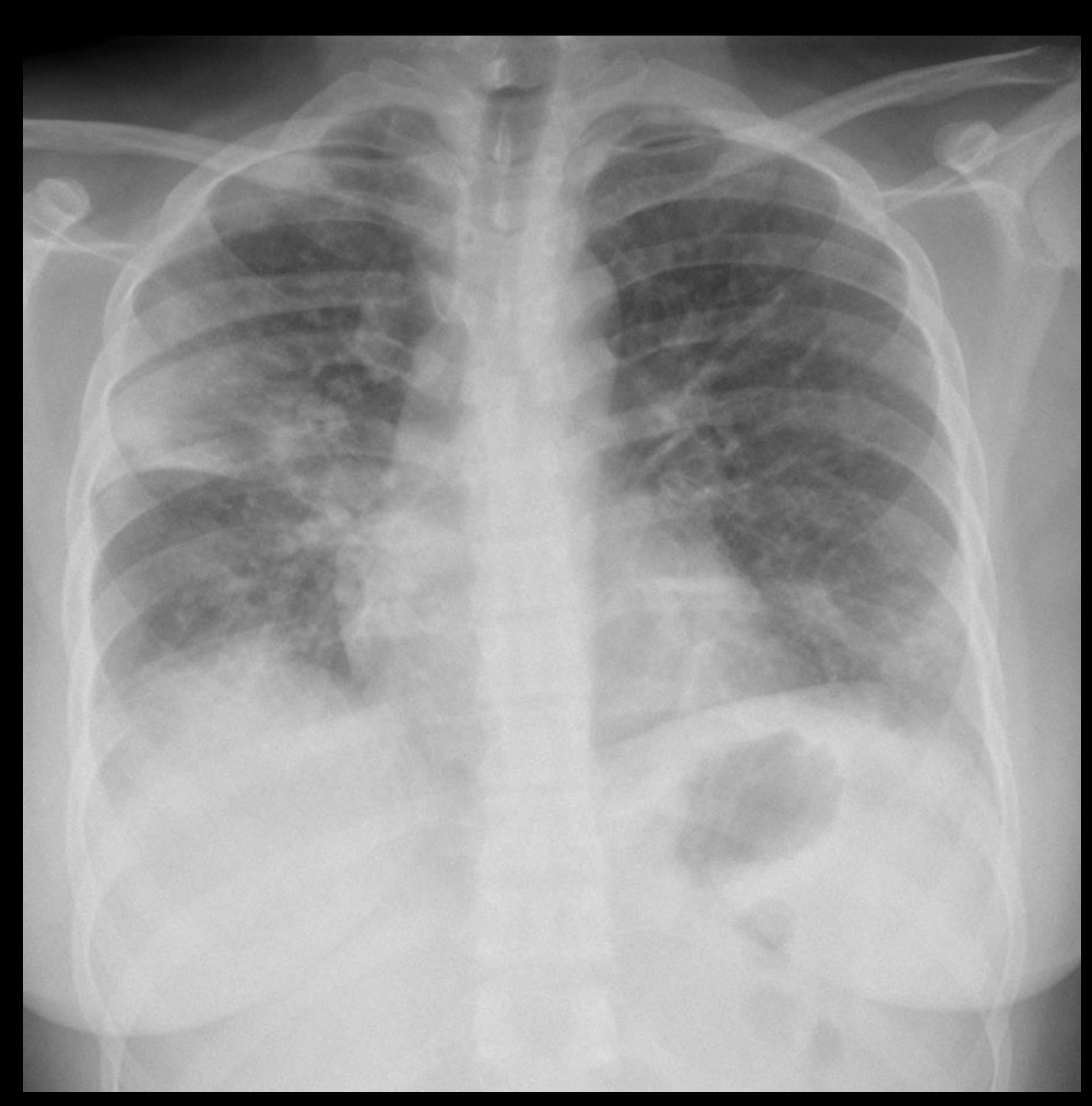


ACUTE LUPUS PNEUMONITIS

AFFECTS ABOUT 4% OF PATIENTS WITH SLE
MORTALITY 50%

SYMPTOMS

FEVER
COUGH
SOB
PLEURITIC PAIN



DIAGNOSIS

LUNG BIOPSY TO DISTINGUISH FROM INFECTION

MAY HAVE A NORMAL CXR!

MANAGEMENT

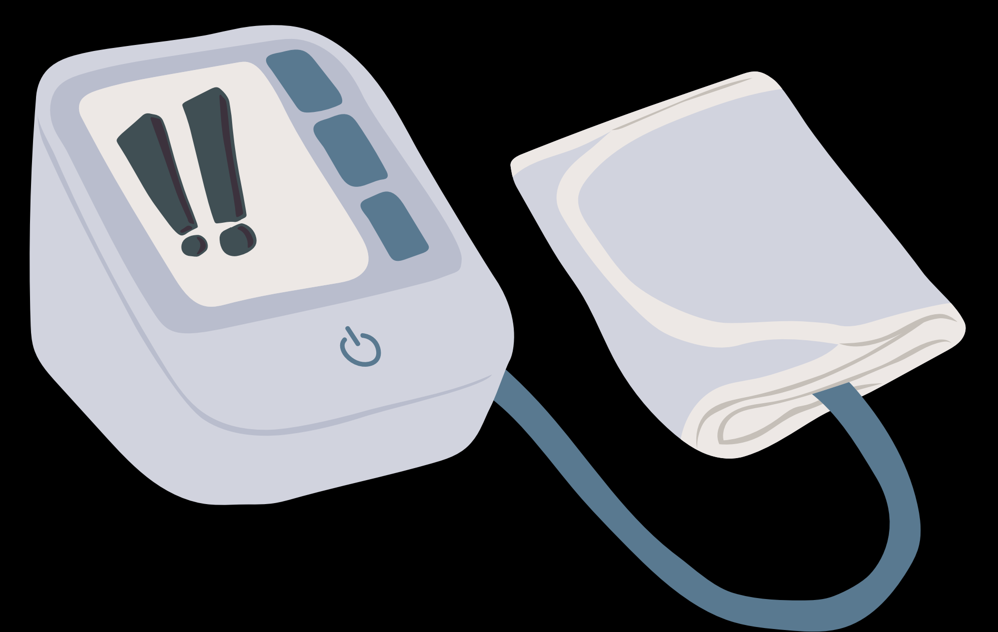
HIGH-DOSE IV PULSE CORTICOSTEROIDS

ANY NEUROLOGIC OR PSYCHIATRIC SYMPTOMS WARRANT CONSIDERATION OF LUPUS AS THE CAUSE

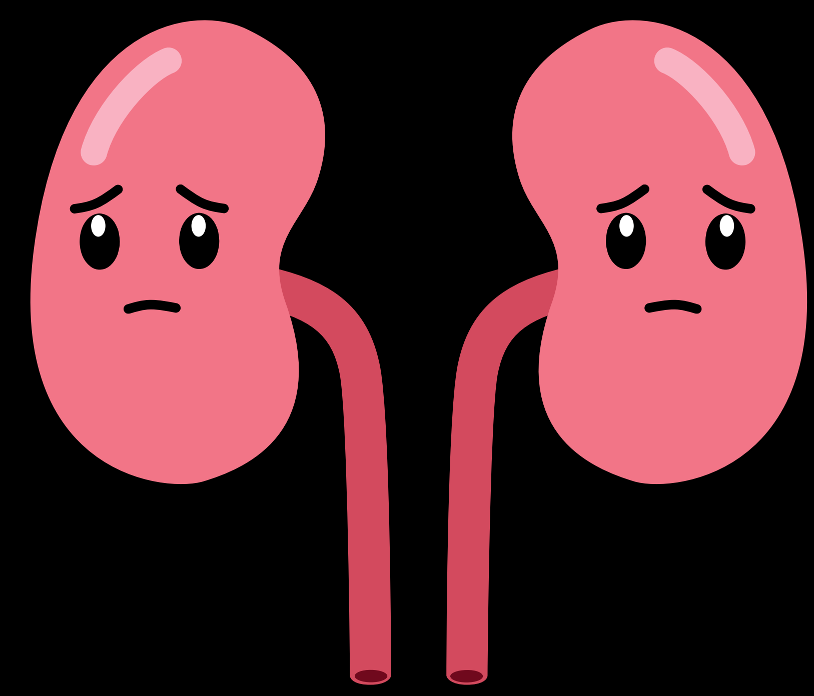
PSYCHOSIS

SCLERODERMA RENAL CRISIS

10% OF PATIENTS WITH DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS



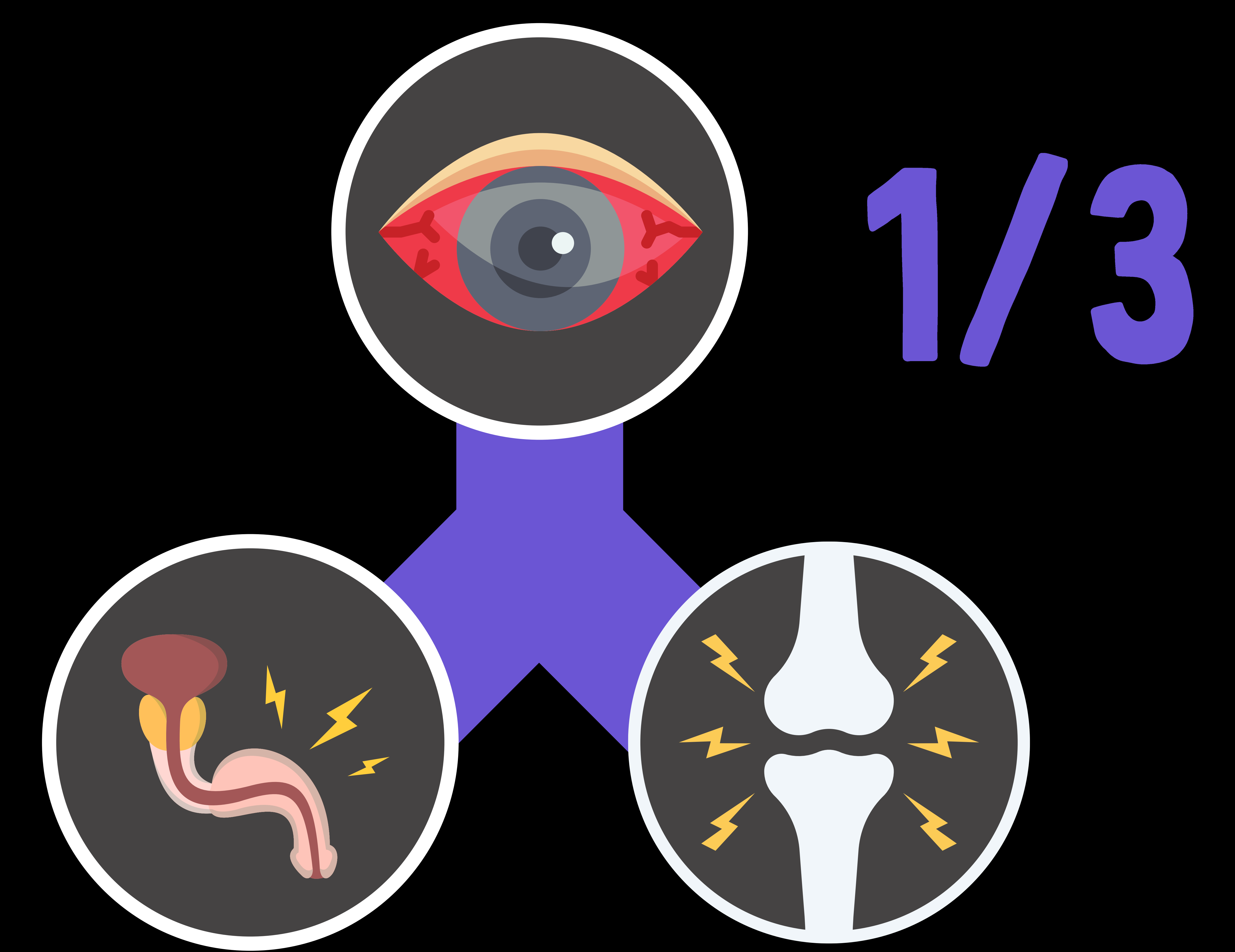
ACUTE ONSET OF SEVERE HYPERTENSION



RENAL INSUFFICIENCY

MICROANGIOPATHIC HEMOLYTIC ANEMIA

TREATMENT: ACE INHIBITORS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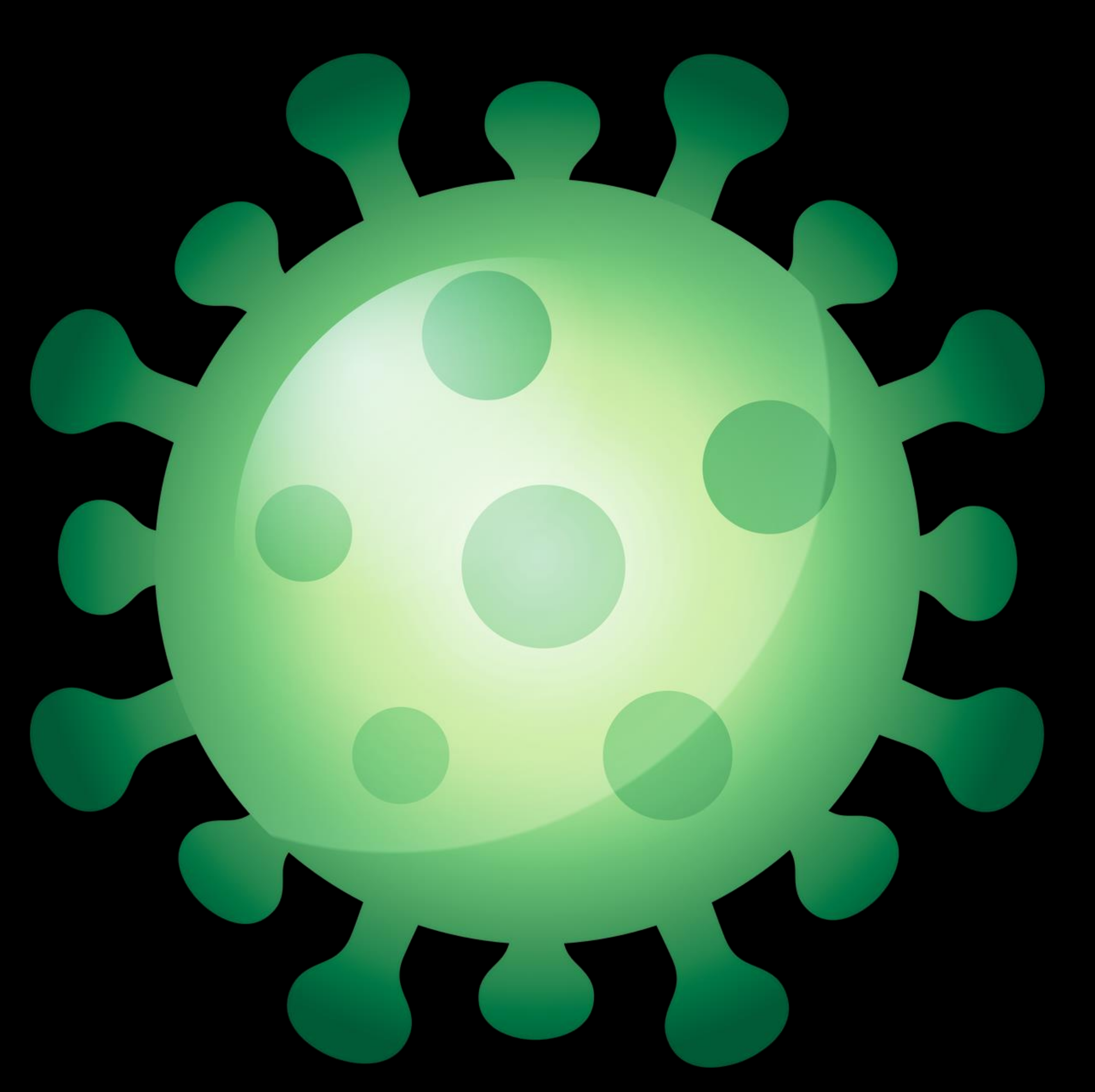
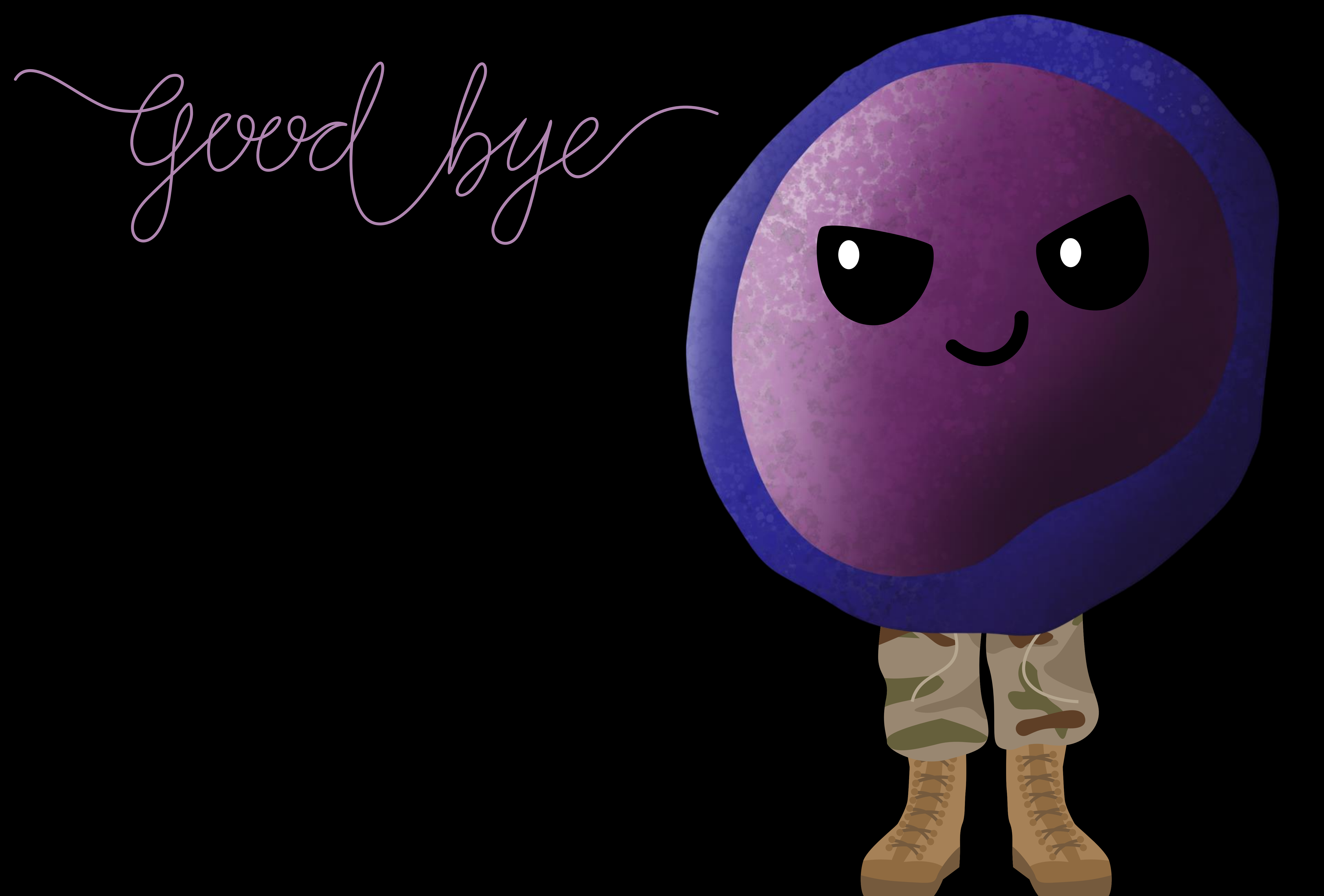


REACTIVE ARTHRITIS IS MUCH MORE COMMON IN PATIENTS WITH

HIV


Good bye