LEPROSY

(Hansen's Disease)

Definition

- Chronic mycobacterial infection
- Peripheral nervous system,skin, other tissues

Naturally acquired leprosy

- Chimpanzee
- Mangaby monkey
- Armadillos

Epidemiology

- *Mycobacterium leprae*

  (variable) 2-7 years

- At Risk population

  Adults (less) 5% conjugal

  Children (more) - 60% in family

PREVALENCE

Total No. of cases - < 3.5 million

Approx. 50 % - IN INDIA

Mode of transmission

- Prolonged intimacy
- Droplet espiratory
- Skin to skin contact
- Tattooing

Bacteriology

M.leprae (1873) - Hansen

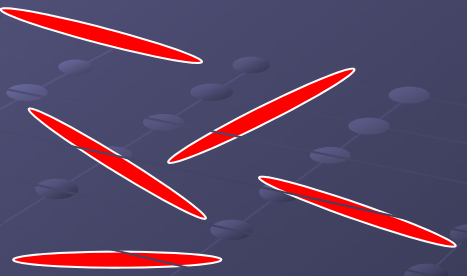
- Non-culturable
- Acid-fast/alcohol fast

(5% H_2SO_4, 3% HCl decolorize)

- Straight – Slightly curved rods
- Rounded ends

  1-8 μm long

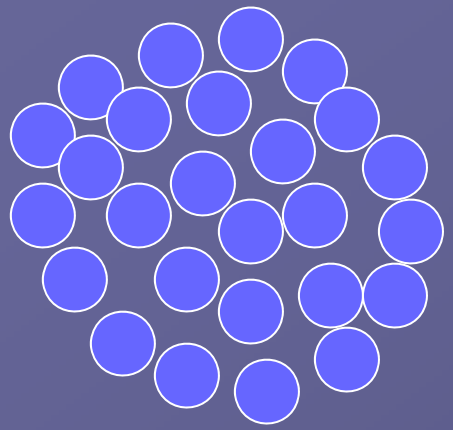
  m diameter



- Obligate intracellular- **GLOBI**

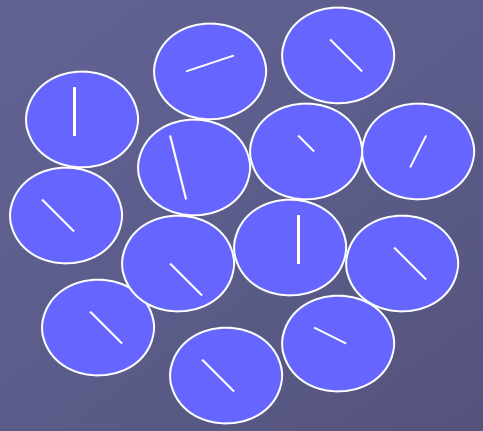
Bacteriological index

- Density of bacilli in high power field



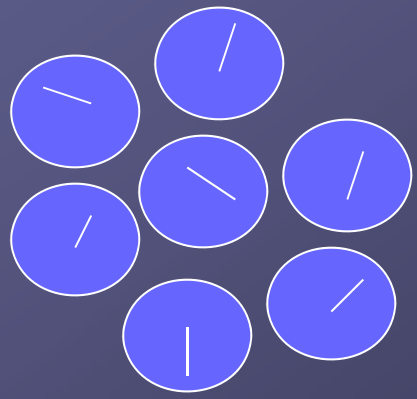
BI-0

No bacilli
in 100 fields



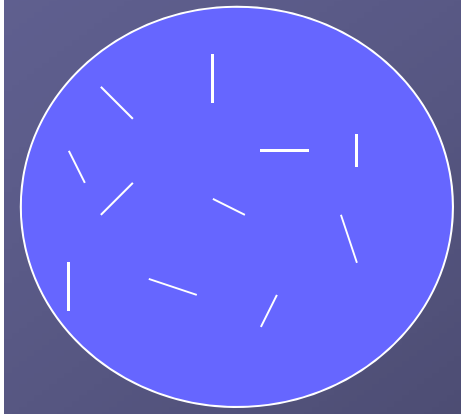
BI-1

1-10 bacilli
in 100 fields



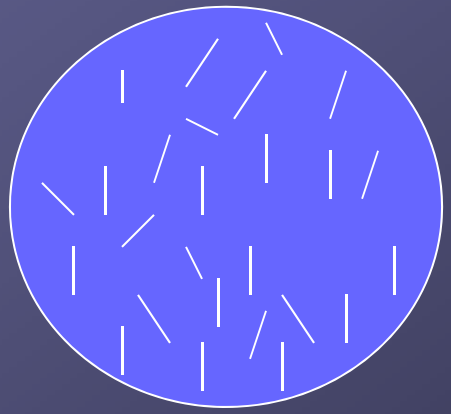
BI-2

1-10 bacilli
in 10 fields



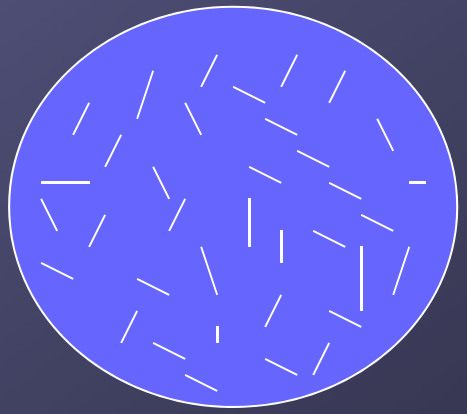
BI-3

1-10 bacilli in
an average field



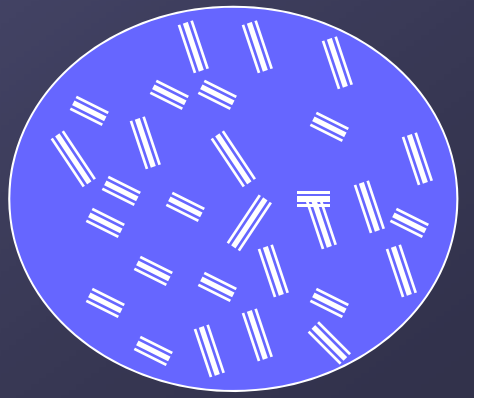
BI-4

10-100 bacilli in
an average field



BI-5

100-1000 bacilli in
an average field



BI- 6

>1000 bacilli in
an average field

Morphological index

- age of presumably live bacilli
- Solid staining - live
- Granular staining - Dead

Culture

- Non-culturable
- Hind foot pad of mice
- Armadillo

PATHOLOGY

Disease of PNS

- Target organ -Schwann cell
- Secondary -Skin (prefers cool areas)

ther organs

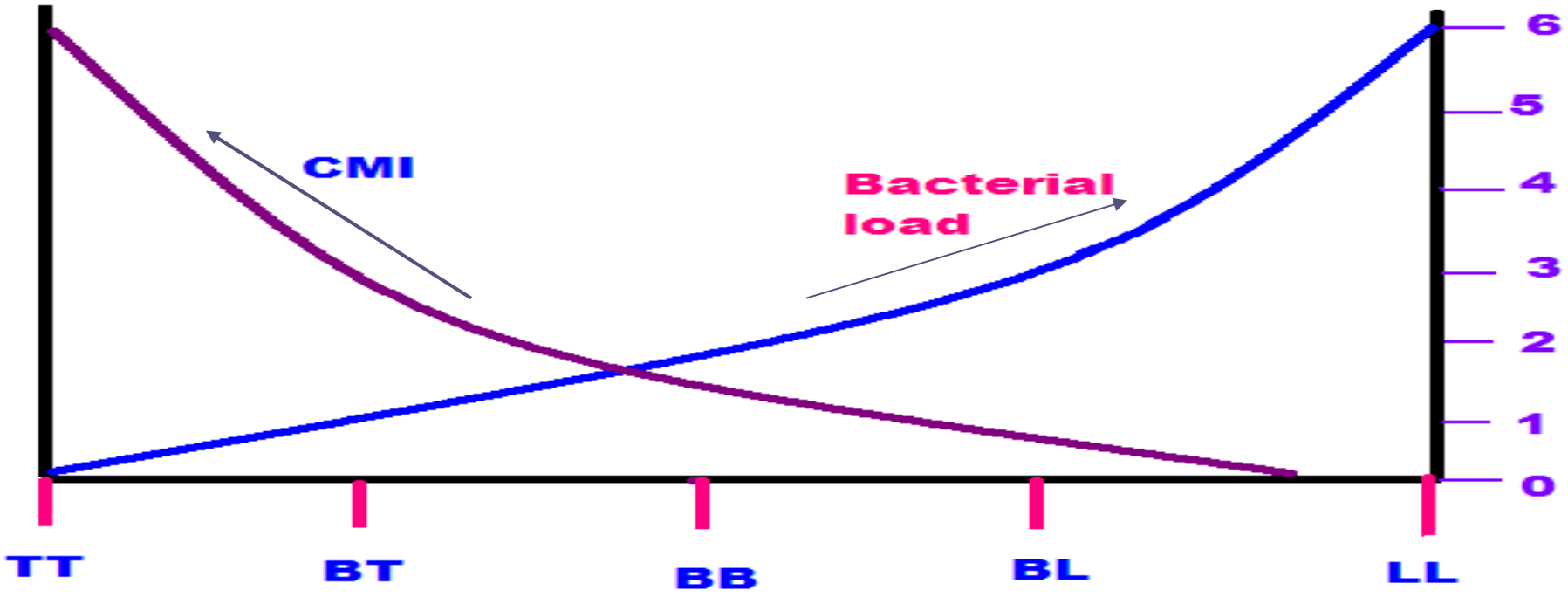
s

- Reticulo-endothelial system
- Nose, pharynx, larynx, trachea
- Endothelium of small blood essels

- Arrector pilorum, dartos
- Skeletal muscles
- Testes

Female reproductive system is spared

- Spinal cord, CNS - **Never involved**
- CMI - AMI – Ig (Nonprotective)



RIDLEY – JOPLING SCALE

Lepromin Test

- **Lepromin A**

  - Autoclaved suspension of Armadillo tissue

-

  - Human tissue

    Non specific

    Not diagnostic

Uses

- To ascertain CMI against M. leprae

  Indeterminate leprosy
- To investigate contacts / high risk
- To classify pure neural disease

Fernandez reaction

Early - 48 hours

Mitsuda reaction

Late - 4-5 weeks

Lepromin

Positive - TT, BT

Negative - BB, BL, LL

Variable - Indeterminate

Classification

RIDLEY JOPLING

TT, BT,BB - Epitheloid cell granuloma

Giant cells

BL, LL Foamy macrophages

Lymphocytes (Lepra cells)

CLINICAL ASPECTS

Tuberculoid leprosy

TT (Localized)

Hypopigmented

Hypo aesthetic/anesthetic

Macule / plaque

Hypopigmented

Hypo aesthetic/anesthetic

Macule / plaque

- Usually single/ (1-3)
- Variable size
- Dry, scaly
- Hair loss
- Absence of sweating
- Absence of triple response

- **Nerve feeding the area**
- Enlarged
- Irregular
- Cord like
- Nodular
- Abscess formation

- **AFB**

  Skin -negative

  Nasal smear -negative
- **Lepromin** -Strongly positive

Secondary changes

- Paralysis of nerve
- Muscular weakness
- Wrist drop
- Foot drop

- Loss of sensation beyond lesions
- Trophic ulcers
- Spontaneous blister
- Osteomyelitis
- Lepra reaction
- Pain (Neuralgia)

BORDERLINE TUBERCULOID (BT)

- 3 -10, variable size
- Hypo- pigmented, Hypo- aesthetic
- Loss of hair, sweat

Skin Absent / scanty

Nasal smear - egative

-

- **AFB**

Skin - Absent / scanty

Nasal smear - Negative

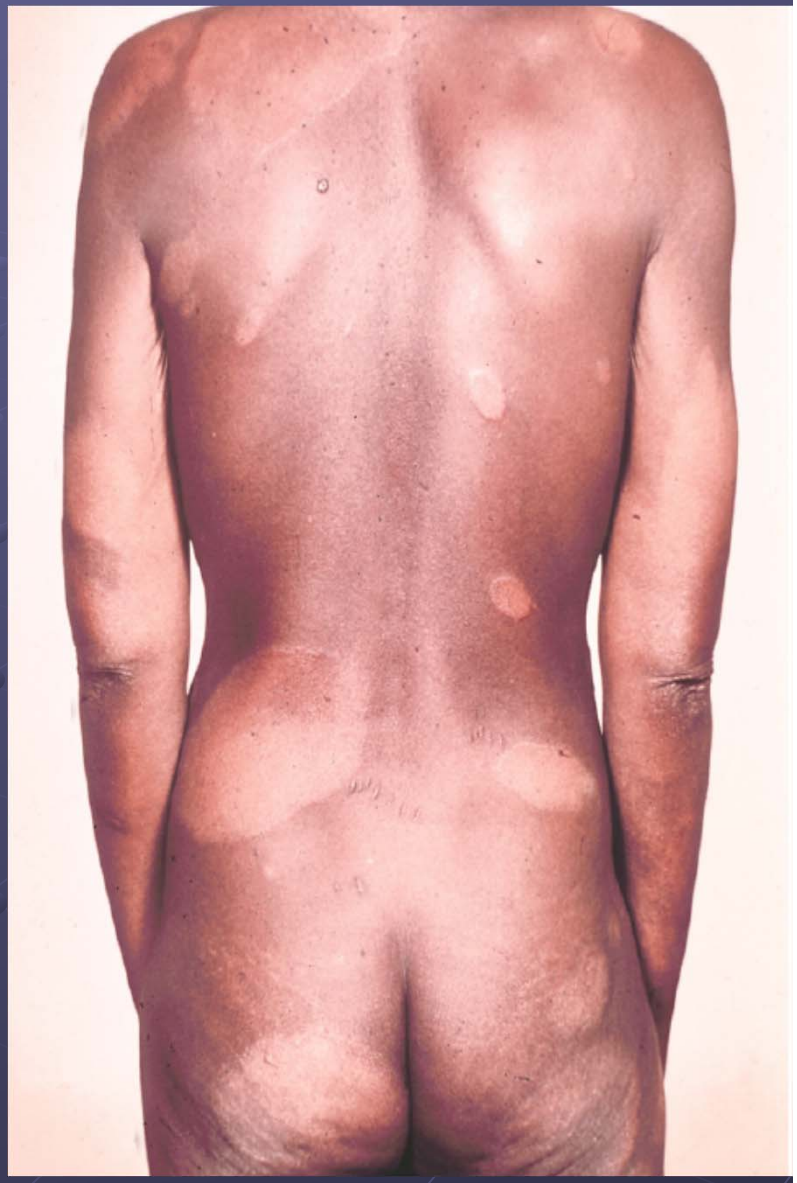
- **Lepromin** - Weakly positive

- **Secondary changes**

  -More common

  - Immunological instability

  - Reactions



MID BORDERLINE(BB)

- Lesions >10

  Variable size

- rythematous shiny plaques
- Loss

SATELLITE LESIONS

Reactions/Secondary changes

- Motor
- Sensory

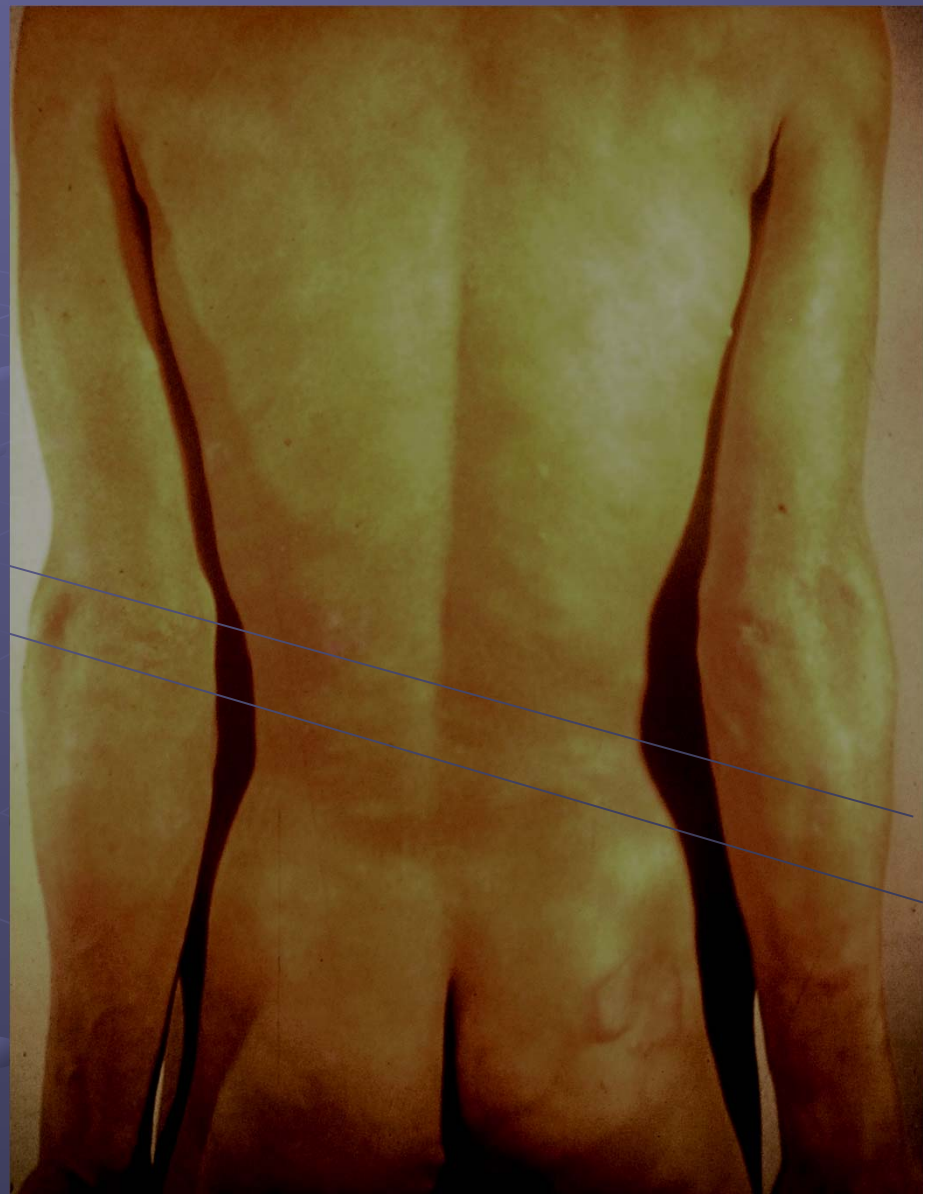
- Punched out lesions (annular)
- Inverted saucer shaped (Swiss cheese)
-

- **AFB**
  - Skin - Moderate
  - Nasal smear - Negative
- Lepromin - Negative

BODERLINE

LEPROMATOUS

- Large no.of lesions
- Papules & nodules
- Plaque (less defined)



fuse areas of infiltration

ce, Hands, feet,Ear lobes

symmetrical thickening of ipheral nerves

ove - stocking anesthesia

me areas of skin still ared


Lp 1

Hair

Minimal loss over lesions

Loss more in nerve distribution

AFB

-Skin -Many

-Nasal -Usually negative

Lepromin - Negative

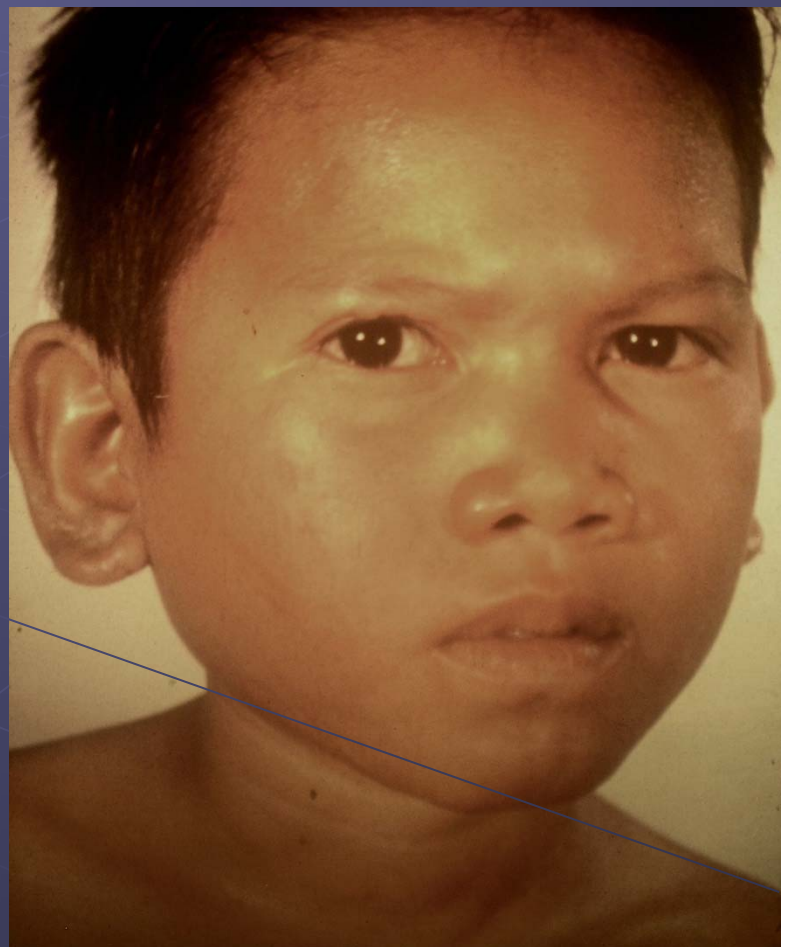
- **Secondary changes**
  - -Reactions
  - -Deformities
  - -Motor palsies
  - -Sensory loss

EPROMATOUS

EPROSY

Diffuse infiltration of skin

Widespread lesions,



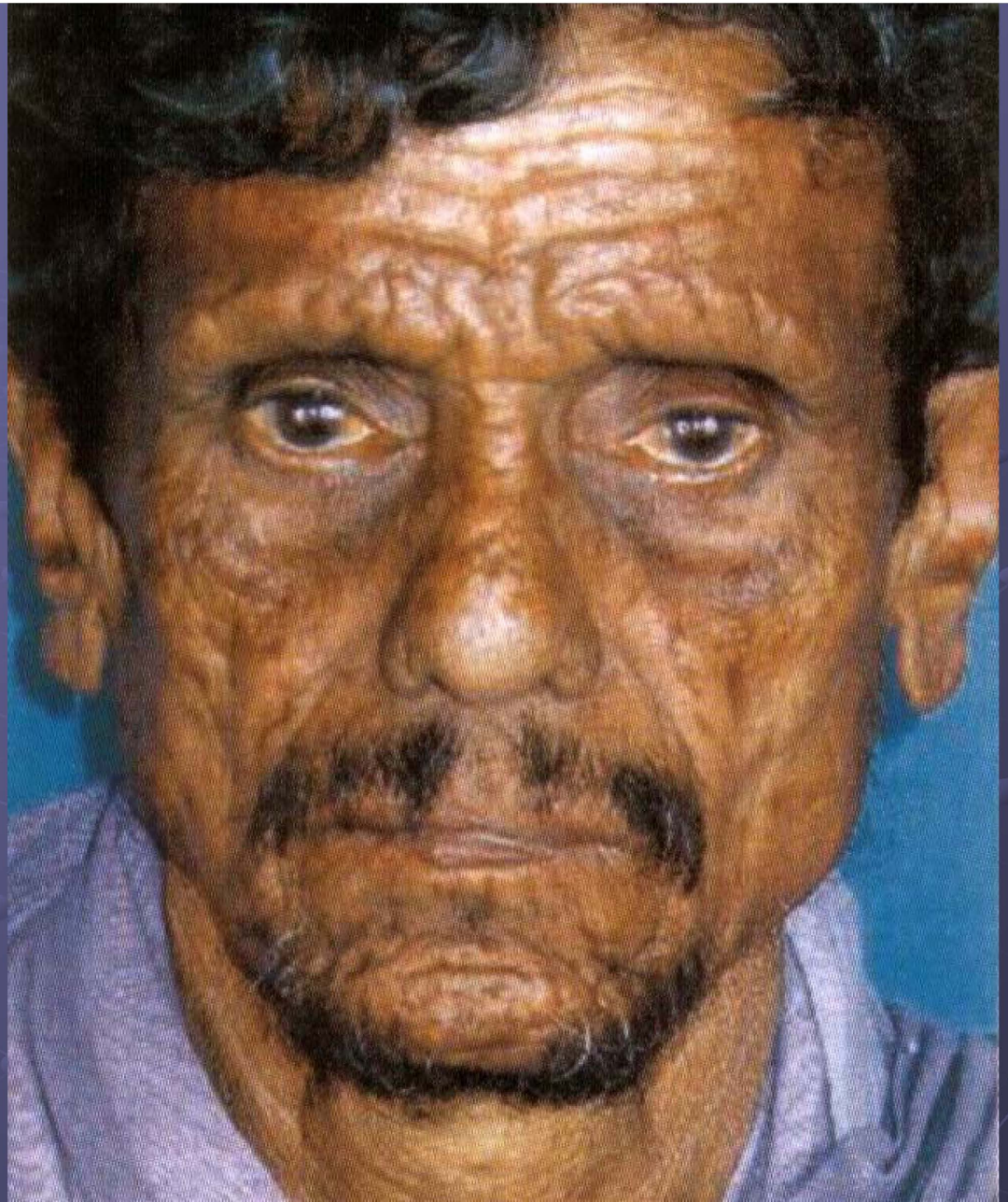
ar lobe infiltration

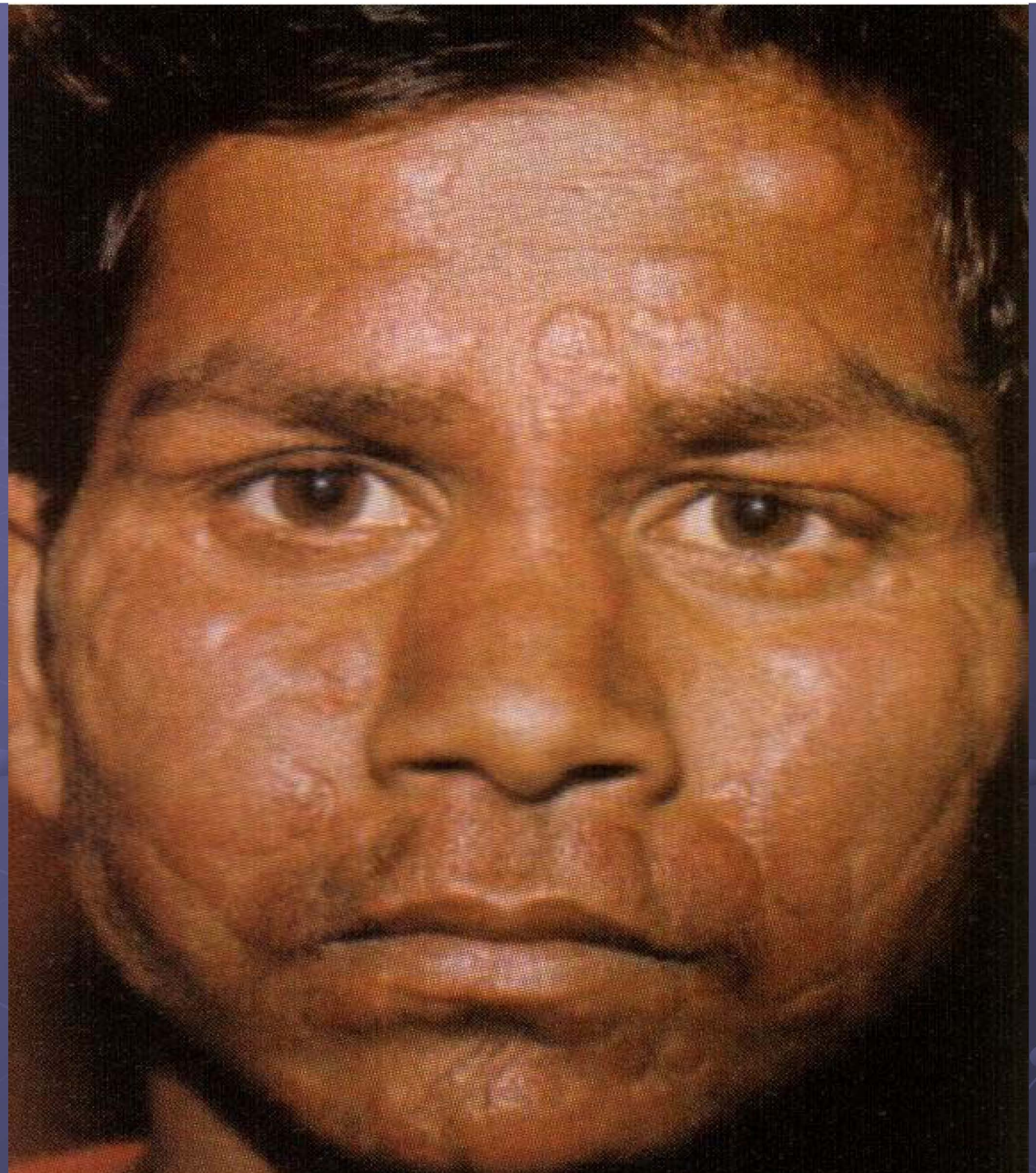
eonine facies

o plaques

apules & nodules
), rarely macules

erves symmetrical
arged





Glove & stocking anaesthesia,icthyosis

B/L madarosis

- Ciliary
- Superciliary

Hair/ sweat - not affected

AFB

-Skin

-Nasal

Globi &clumps 6 (+)

Lepromin - Negative

YSTEMIC FEATURES

- Epistaxis,nasal stuffiness,saddle nose
- B/L pedal edema
- Eyes - Iridocyclitis
  -
- Bones
  eriostitis(Hand/Feet/Skull)

Testes - Atrophy

- Acute epididymo-orchitis

Kidney - Amyloidosis,

Glomerulonephritis

Lymphadenopathy

Lepra reaction type II

NERVE INVOLVEMENT IN LEPROSY

ERIPHERAL NERVES

Supraorbital & supra trochlear N

Infraorbital & infratrochlear N.

Supra clavicular N.

- Lateral,Middle,Medial

Greater auricular N

Radial N. & Radial cutaneous Br.

Ulnar N

Median N

Lateral,medial & intermediate

cutaneous N. of thigh

Lateral popliteal N

Post tibial N

CRANIAL NERVES

Olfactory nerve

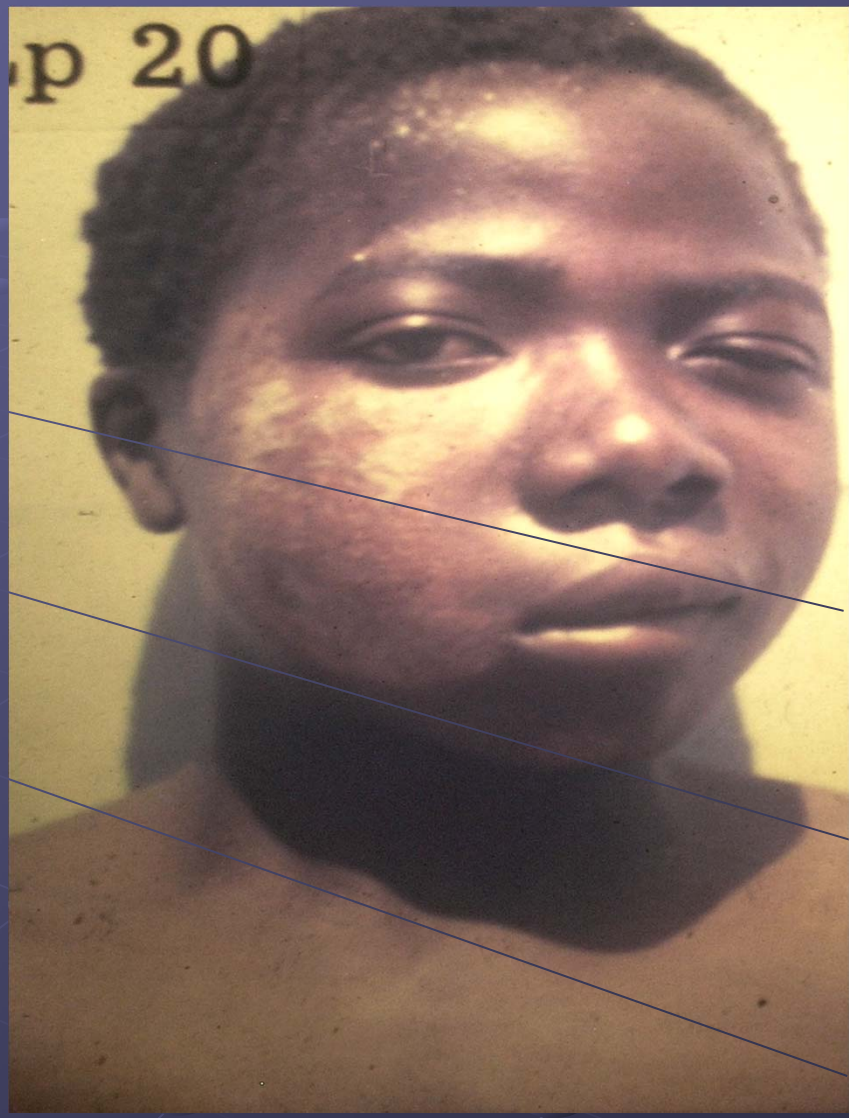
Fifth nerve

Seventh nerve(Facial)

- Zygomatic branch

acial N palsy

- LMNP
- Most commonly
- with lesion over face
- Lepra reaction
- Bilateral in few

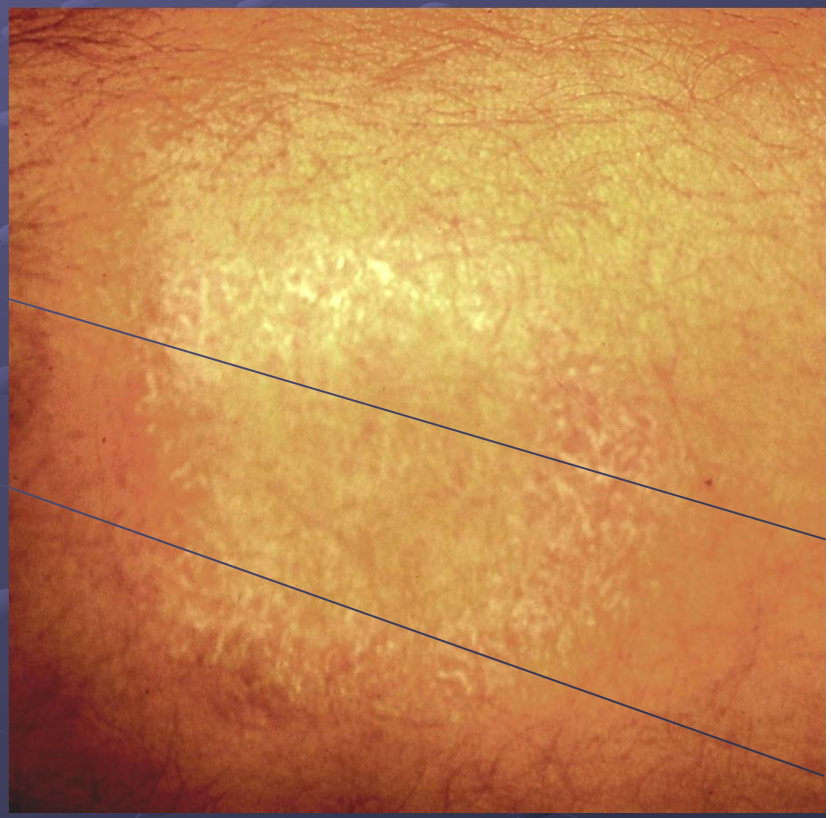


NDETERMINATE

EPROSY

mmunity

- Not defined
- Few hypo-pigmented macules
- Anesthesia ±
- Nerves ±



- Smear -ve
- May heal spontaneously
- May evolve as BB, BL, LL, BT
- Lepromin - Variable

LEPRA RACTIONS

Hypersensitivity states

Type I - Delayed type hypersensitivity (CMI)

Type II - Immune complex mediated (ENL)

TYPE I LEPRA REACTION

Without treatment

TT→BT → BB → BL → LL

With treatment

- Regain of CMI
- On treatment -about 6 months
- BB, BT

- **Clinical Features**
  - Swelling, erythema, ulceration
    Lesions
    Acute Neuritis
  - Paralysis, Neuralgia
  - Fever/malaise
    - Rarely

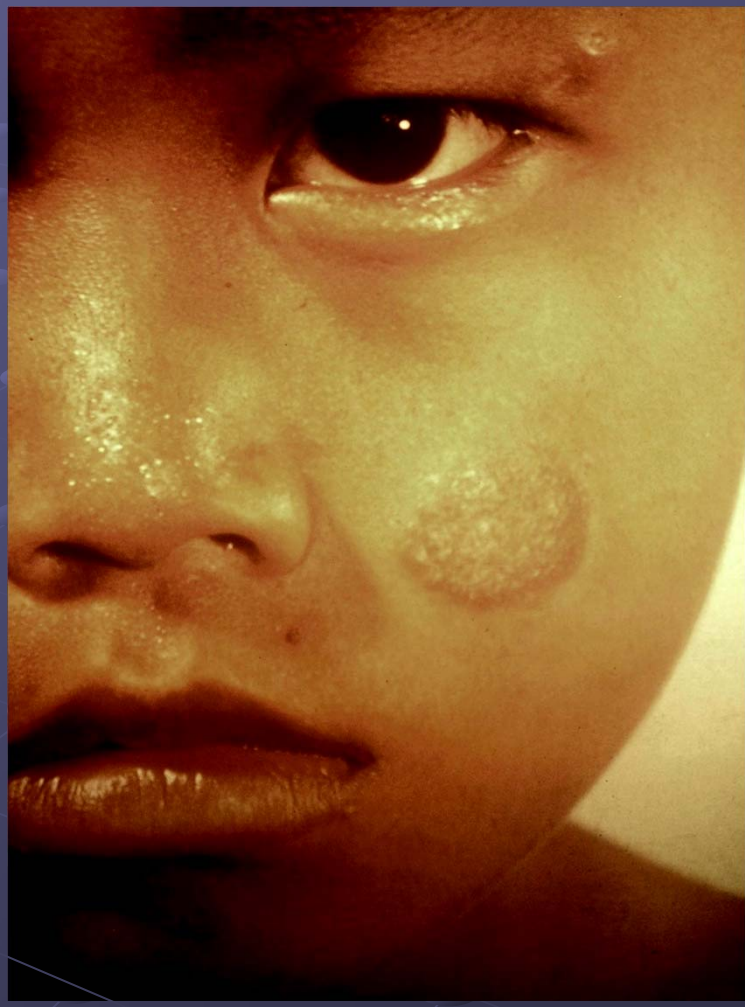
Treatment

Lepra I

- Mild - NSAIDS

  Severe

  - Systemic steroids
  - Glucocorticoids
  - Surgical intervention



TYPE II LEPRA REACTION

Erythema Nodosum Leprosum (ENL)

Immune complex deposition

- LL,BL
- No relation with treatment
- May be presenting symptom

linical features

Multiple system Involvement

kin

ENL : **VASCULITIS**

Bright erythematous,

tender, papule, nodule

plaques

- Eye : Acute iridocyclitis
- Rhinitis, epistaxis
- Ac. Lymphadenitis - generalized
- Ac. Epididymo orchitis

- Ac. Periostitis - Bone pains - tibia
- Ac. Myositis
- Ac. Glomerulonephritis
  - Proteinuria
  - Casts
- Ac. neuritis

Factors precipitating Type 2 reaction

- Inter-current illness /Infection
- Injury, surgical intervention
- Stress -Physical / mental
- Montoux test - vaccinations
- Pregnancy - delivery

TREATMENT

- NSAIDS
- Glucocorticoids - 1mg/kg body wt.
- Thalidomide - 100mg TDS
- Clofazamine - 100mg TDS
- Colchicine 1.5-2.0mg daily
- Chloroquine - 250mg TDS

Diagnosis of Leprosy

Investigations for diagnosis

- Slit skin smear
  - SSS
  - BI
  - MI
- Nasal smear

- Skin biopsy
- Nerve biopsy
- Histamine test axon reflex
- Sweating test
- Lepromin test

For treatment & follow up

- HMG,LFT,RFT
- Urine R/E
- X-ray chest
- G-6-PD deficiency (if suspected)

OTHERS

- Normocytic / Normochromic anaemia
- Biological false positive VDRL
- Rh factor, ANF, LE cells
- ESR
- Thyroglobulin Ab
- Anti spermatozoal Ab, Cryoproteins

TREATMENT OF LEPROSY

ANTILEPROSY DRUGS

First line drugs

Dapsone

Rifampicin

Clofazamine

Others :

Ethionamide

Thiacetazone

DAPSONE(DDS)

Diamino–diphenyl sulfone

- Dihydrofolate synthetase inhibitor
- Bacteriostatic

- 6-10mg/kg body wt./week

  100mg OD >50kg

  50 mg OD <50kg

  Half life - 28 hours

- Cheapest Most effective,

  safe in pregnancy

SIDE EFFECTS

- Light headedness/headache
- Hemolytic anemia
- Agranulocytosis
- Hepatitis

- Psychosis
- Fixed drug eruption
- Exfoliative dermatitis
- Methemoglobinemia

CLOFAZIMINE

- Red iminophenazine dye
- Bacteriostatic
- Mechanism - unknown
- Anti-inflammatory action

- Anti leprosy dose - 50mg/OD

  Anti inflammatory dose -100mg TDS
- Half life - 70 days

SIDE EFFECTS

- Reversible dose-related, reddish brown discoloration of skin, hair,sputum, urine, faeces
- Keratoderma - Acquired ichthyosis
- Phototoxicity, acneiform eruption

- Abdominal syndrome

  Early (1 month) :Diarrhoea/abdominal pain

  Late (>3months): Diarrhoea /abdominal pain

  - Loss of weight
  - Death
  - Deposition of CLOFAZIMINE CRYSTALS

RIFAMPICIN

- Potent bactericidal
- Inhibits RNA synthesis (DNA dependent RNA polymerase)
- Dose-10mg/kg body weight
  - Maximum 600mg/day
  - 450mg <35 kg
- MI - 0 (5 weeks)

SIDE EFFECTS

- Red discoloration of urine

  Intermittent administration.

  a) FLU Syndrome -Fever, chills,rigors,bone/ joint pains, malaise,headache

  b) Syndrome of shock, renal failure, hemolytic anemia, purpura

OTHERS

- Hepatitis
- Thrombocytopenia
- Psychosis
- Osteomalacia

- Pemphigus vulgaris
- Steven Johnsons syndrome
- Porphyria cutanea tarda

Drug interaction

-Induction of hepatic microsomal enzymes

(Steroids /oral contraceptive)

TREATMENT REGIMEN

Classification for treatment purposes

Paucibacillary-Smear negative

(TT,BT)

Multibacillary -Smear positive

(BB,BL,LL)

MULTI DRUG THERAPY : (MDT)

WHO

Need : Drug Resistance - Dapsone

WHO MDT (PB)

Dapsone 100mg OD (Self administered)

Rifampicin 600mg once monthly

(Supervised)

Duration - 6 months

Children (0-5year)

- Dapsone 25mg
- Rifampicin 300mg

(6-14 year)

- Dapsone 50mg
- Rifampicin 450mg

WHO MDT (MB)

- Dapsone -100mg OD (Self administered)
- Rifampicin-600mg once monthly (supervised)

Clofazamine-50mg OD (self administered)

300mg once monthly (supervised)

Duration - 2 years

NLEP MDT

PB - Same as WHO

MB - Intensive phase + WHO

tensive phase

2 weeks daily - supervised

Rifampicin - 600mg

Clofazimine - 100mg

Dapsone - 100mg

x 14 days

Newer drugs

- Clarithromycin
- Ofloxacin
- Minocycline
- Rifabutin
- Pefloxacin
- Dapsone analogues

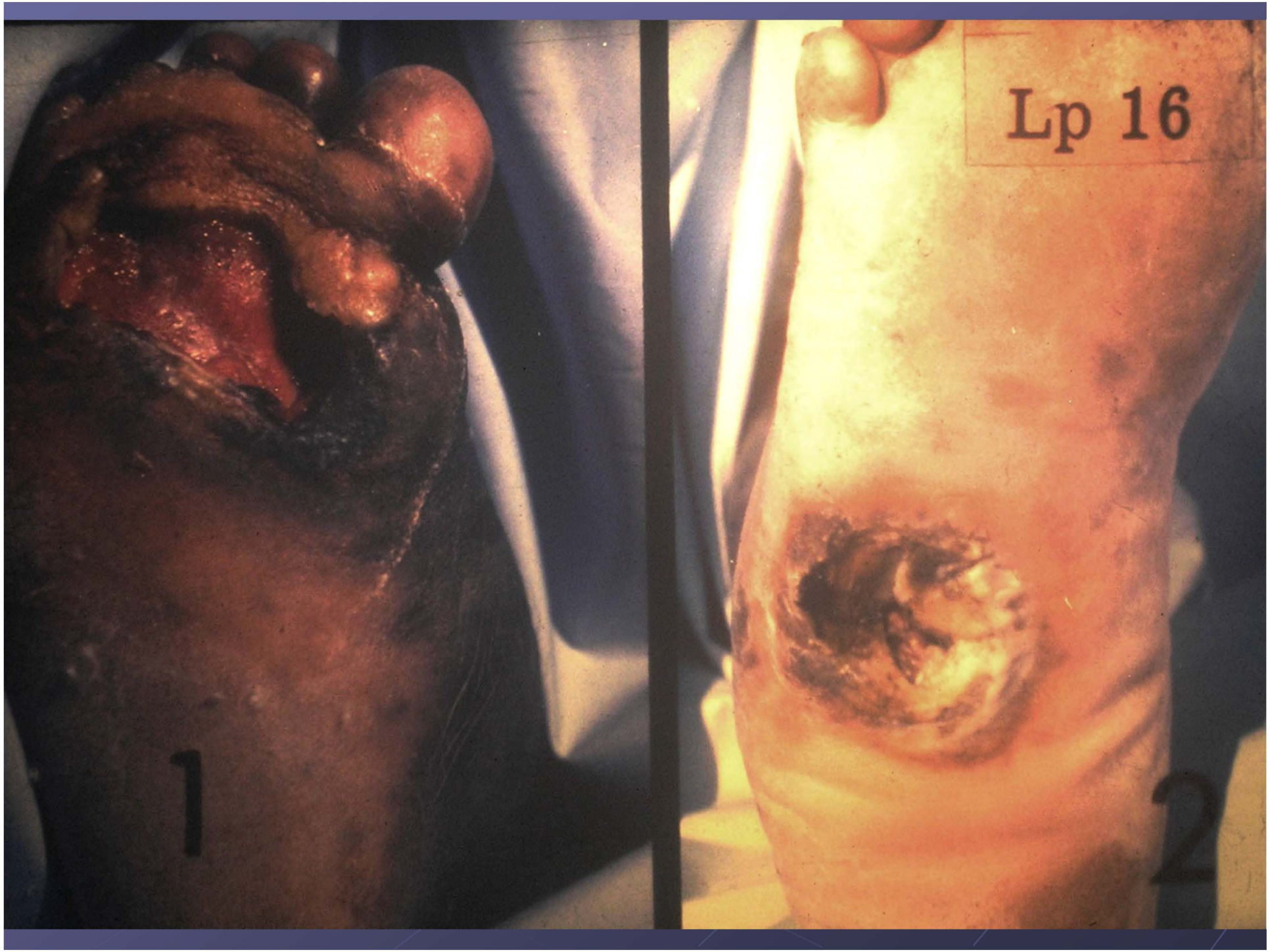
REHABILITATION

- Cosmetic correction
- Profession adjustment

REHABILITATION

- Cosmetic correction
- Profession adjustment


A
B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