HANSEN'S DISEASE

By Stephanie Burns, RN, DNC

Objectives

- To discuss brief history of Hansen's disease
- To discuss how to diagnose Hansen's disease
- To explain classifications of Hansen's disease
- To discuss reactions in Hansen's disease
- To discuss treatment of Hansen's disease

HANSEN'S DISEASE

HISTORY

- 1.) Leprosy is a chronic infectious disease caused by Mycobacterium leprea bacteria.
- 2.) The earliest descriptions recorded about 600 BC in India.
- 3.) 1873 Gerhard Hansen, a Norwegian physician, was recognized as first person to identify the bacterium, Mycobacterium leprae, as the cause of the disease.
- 4.) During this period until early 1900's leprosy was treated with hydrocarpus oil. This oil was extracted from dried fruit of the hydrocarpus tree.

- 5.) Leper colonies have existed since the Middle Ages with purpose to isolate lepers and prevent spreading the disease.
- 6.) 1941 sulfone, (promin), was introduced to treat HD. Although this medication is reported as being somewhat successful in curing HD, it was administer IM and quite painful.

- 7.) 1950 dapsone became the standard treatment for HD
- 8.) 1982 The World Health Organization recommended multidrug therapy (MDT) with dapsone, rifampin, and clofazimine.

- 1995 WHO has made MDT available free of charge to all patients worldwide

DEFINITION OF PREVALENCE OF LEPROSY

- Refers to estimated population of people who are managing Leprosy at a given time

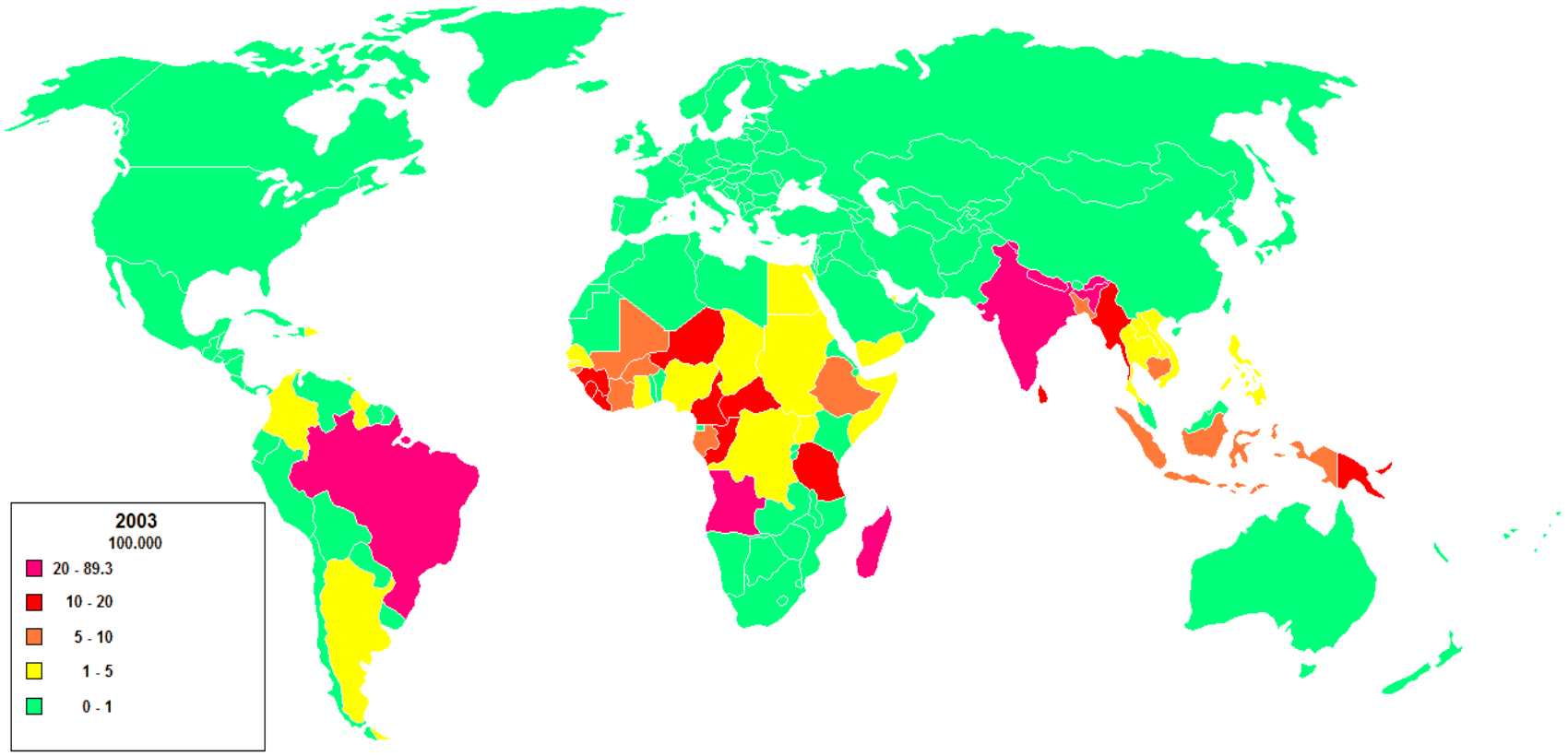
DEFINITION OF INCIDENCE OF LEPROSY

- Refers to the annual diagnosis rate or the number of new cases of Leprosy diagnosed each year

- Currently Brazil, Madagascar, Mozambique, Tanzania and Nepal are listed as having 90% of the Hansen's disease cases.

Beginning of 2008 – Global registered prevalence of leprosy = 212,802. New cases detected during 2007 was 254,525

2011 - global prevalence registered at 192,246 cases. New cases detected during 2010 was 228,474 (excludes small # of cases in Europe.


2003
100.000
20 - 89.3
10 - 20
5 - 10
1 - 5
0 - 1

U.S. DATA

- 6500 cases of Leprosy in U.S.
- 90% immigrants from countries where disease is endemic.
- 600 cases with active disease and requiring drug treatment
- 200 – 250 cases reported each year.

U.S DATA CONTINUED

- The Largest #’s of Leprosy cases in U.S. are in
  - California
  - Texas
  - Hawaii
  - Louisiana
  - Florida
  - New York
  - Puerto Rico

LAHEY CLINIC DATA

	2010	2011	2012
Pts on Roster	269	272	276
Newly Dx	6	4	3
Active Status	95	99	102
Brazilian Pts	6	4	2

REASONS FOR DECREASE IN REGISTEREDED CASES

- Shortened treatment period and revised definition of a case of HD. Current definition of a case, according to WHO is a patient requiring chemotherapy.

- Therefore, patients who have completed their chemotherapy are no longer counted as cases.

- In summary the number of cases on treatment has decreased over the last decade but the new cases have not dramatically decreased

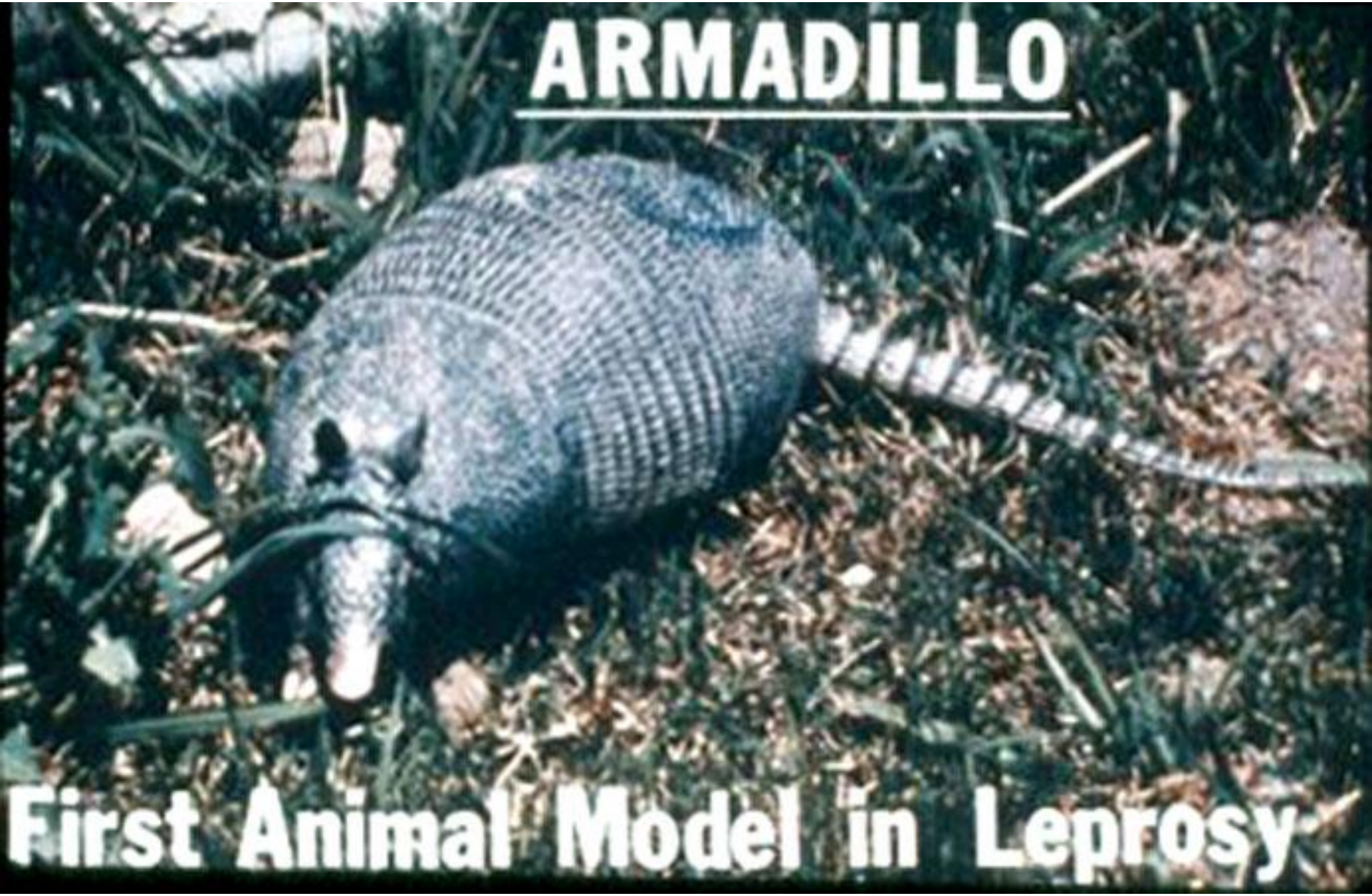
RISK FACTORS

- Highest risk for population living in endemic areas with poor living conditions, contaminated water, poor diet.
- Individuals with compromised immune system
- Men are two times more likely to contract leprosy than women

- Usual source of infection is from patients with untreated lepromatous HD. Large amounts of bacilli can be found in their upper respiratory tract and nasal mucus membranes.
- More prevalent with prolonged and/or close contact with patient.

- Transmission of M leprae is through droplet or dust.
- M leprae can remain viable,outside the body, for 24 hours to several days.
- There is no evidence of sexual transmission.

- Armadillos found in Texas and Louisiana, have been found to be infected with M leprae. Therefore are suspected of being a source of transmission of the disease to humans.


ARMADILLO
First Animal Model in Leprosy

- Heredity theory effecting the immune system. Some family members have predisposition to disease. (Example:Why one sibling exposed to the disease will become infected, while another sibling having same exposure will not contract HD)
- Incubation period once infected by m leprae can be from 6 months to 40 yrs.

DIAGNOSIS

- THREE CARDINAL SIGNS

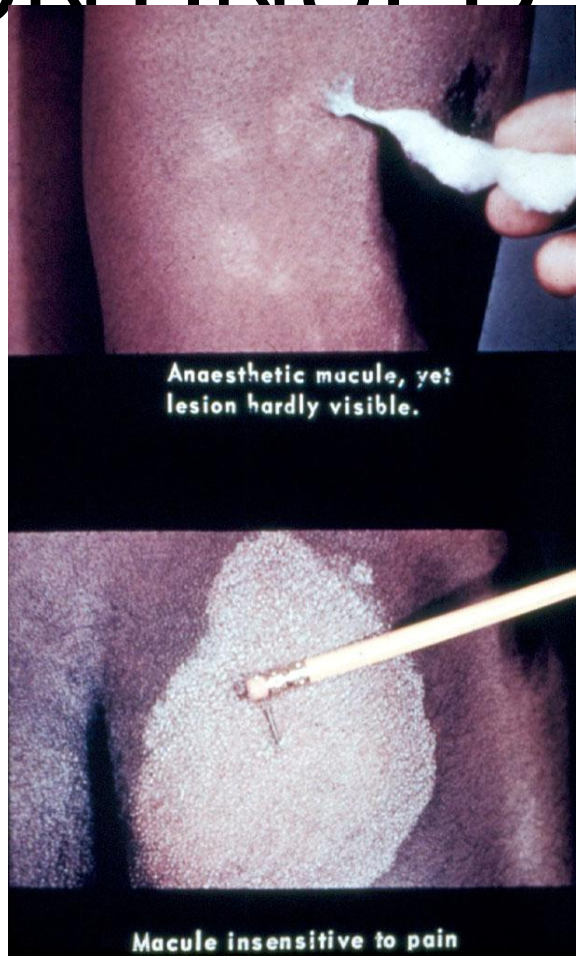
1.) Anesthesia

2.) Enlarged nerves

3.) Acid-fast bacilli

DIAGNOSIS CONTINUED

- Localized lesion
  - Hypopigmented
  - Hyperpigmented
  - Erythematous
  - Sensation loss

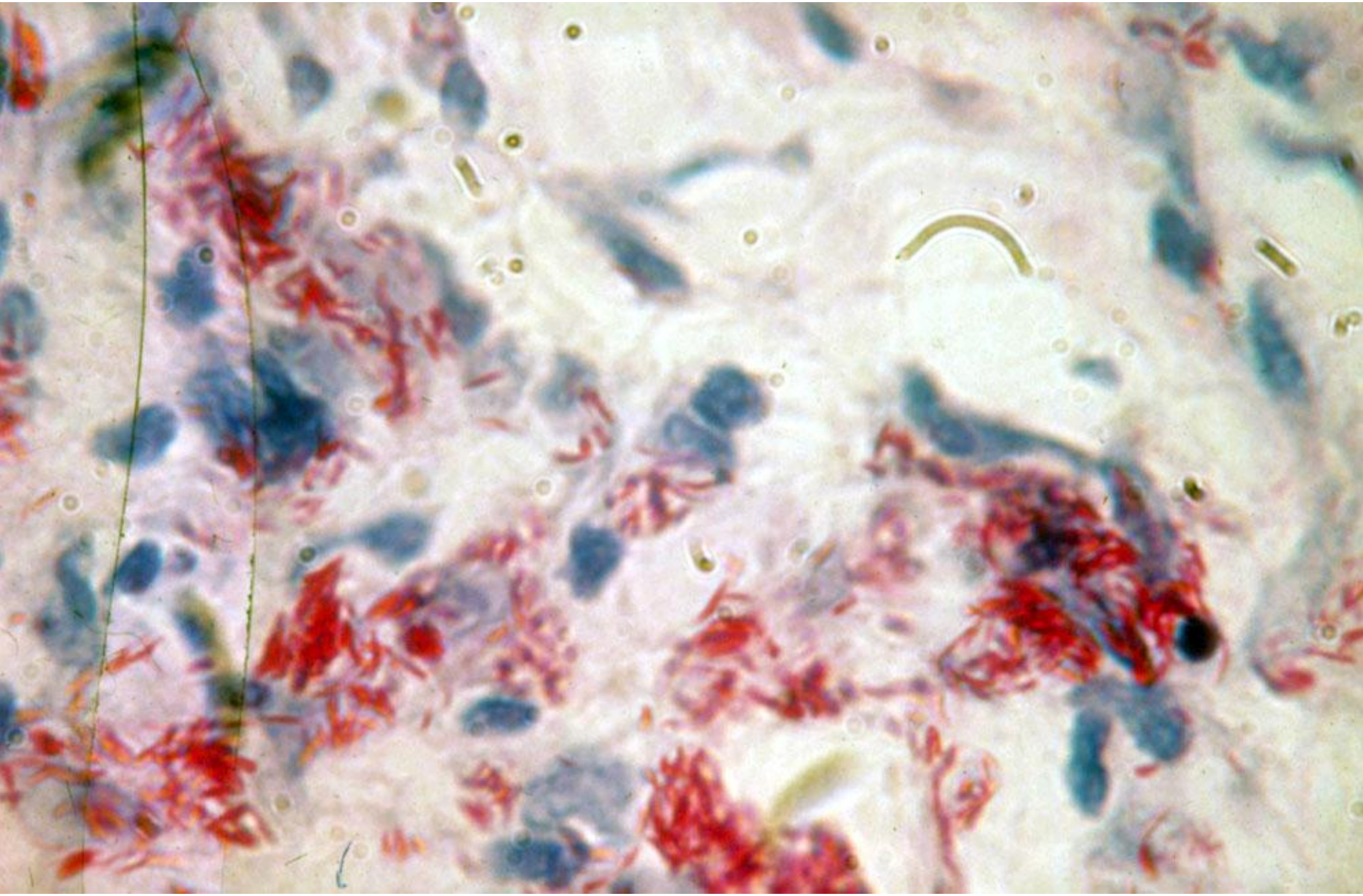


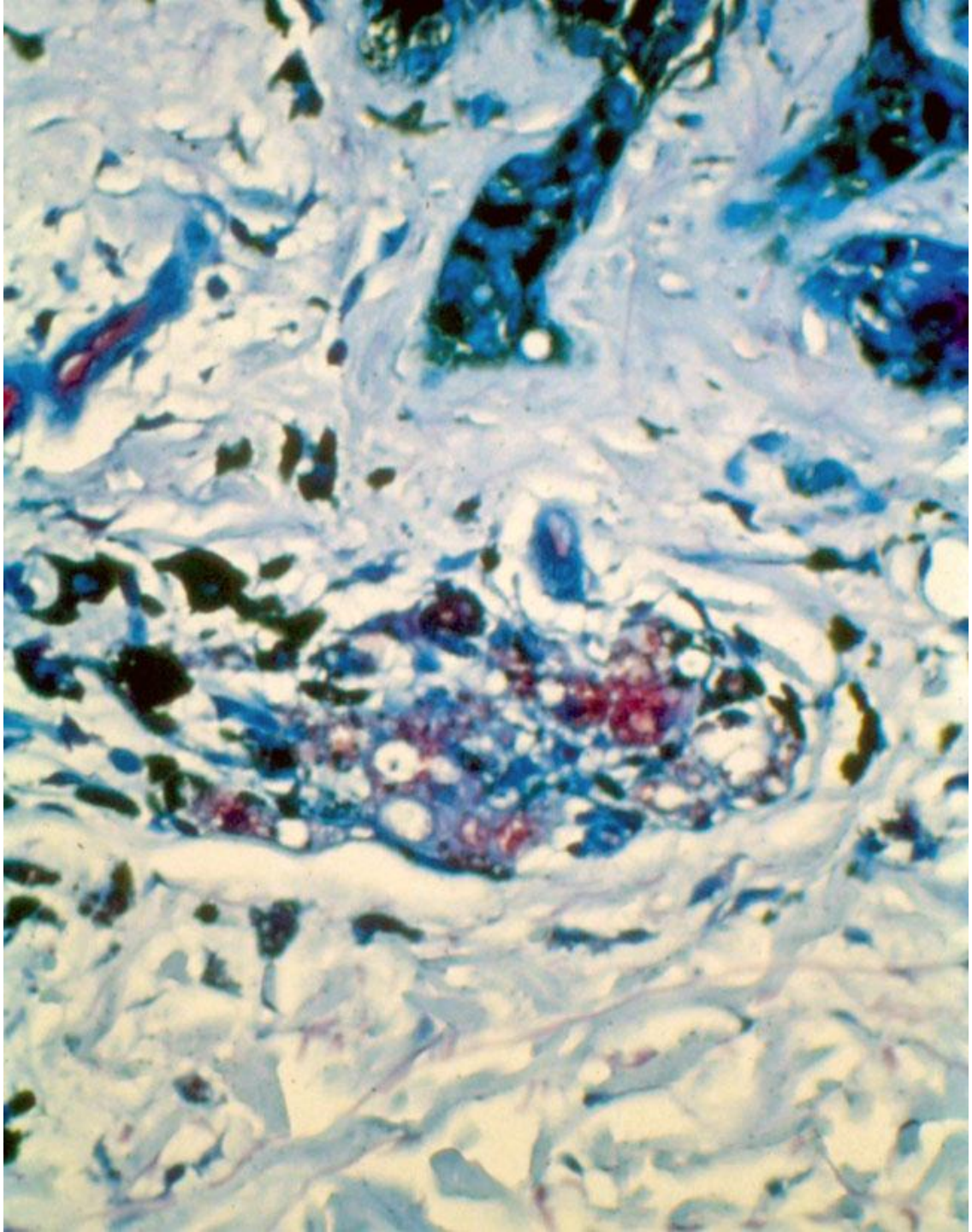
- Nerve involvement is always present in HD
- Bacilli can be found in nerves at almost any level from periferal nerves to the larger proximal nerve trunks.

- Acid bacilli can be detected in skin through:

  skin smears

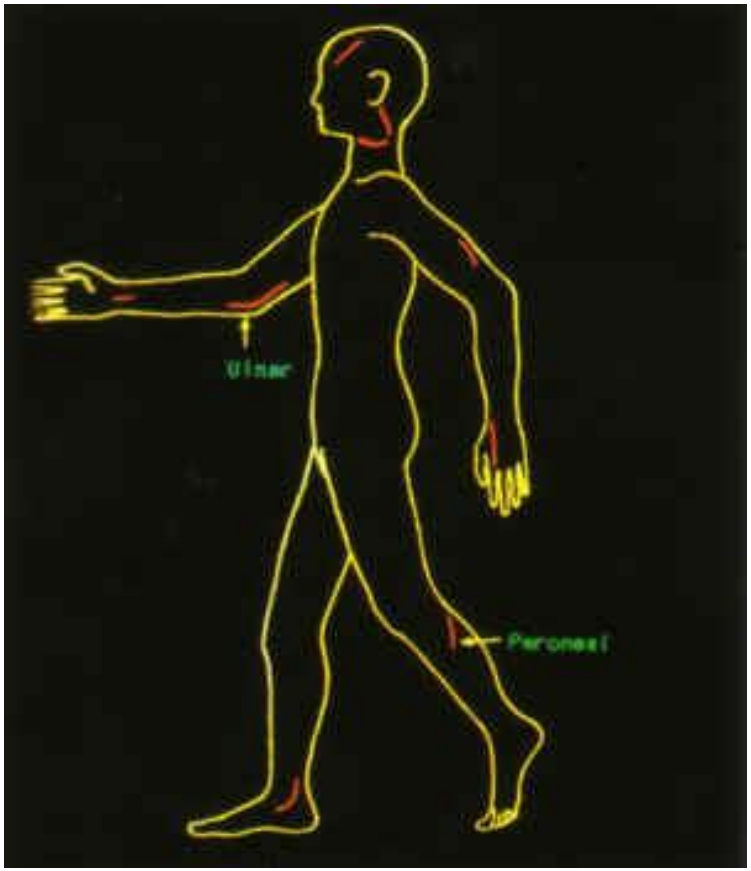
  skin bx





PERIPHERAL NERVES

- FACIAL
- GREAT AURICULAR
- MEDIAN
- ULNAR
- RADIAL
- RADIAL CUTANEOUS
- PERONEAL
- POST TIBIAL



- Bacilli tend to accumulate in the cooler parts of the body
  - Ears
  - Forehead
  - Chin
  - Nose
  - Eye
  - Elbow
  - Wrist
  - Knee
  - Testicles
  - Ankle

- Nerves are usually superficial, enlarged, painful and can be palpated easily.
- Damage to the nerves cause sensory, and motor damage

Examination of the skin, eyes, hands, and feet.

- Skin - Skin lesions should be tested with monofilament or article that is able to give a light touch, such as a cotton wisp or feather. This test will indicate if patient has anesthesia within skin lesions.
- Eyes – look for signs of redness, tenderness or evidence of lid weakness, which can cause drooping of eye lid (lagophthalmus).

DIAGNOSIS CONTINUED

- Hands – observe for loss of sensation, weakness of the intrinsic muscles of the hand or muscle atrophy which can cause claw hand deformity

- Lower extremities – examine for loss of sensation, muscle atrophy, foot drop, or claw toes. Due to insensitive feet always check for pressure or trauma ulcers.

CLASSIFICATION OF HANSEN'S DISEASE

- Two systems of classification
  - 1.)Ridley and Jopling classification
    - » Clinical
    - » Immunology
    - » Histology
    - » Bacteriology
  - 2.) the WHO two group classification
    - » Pausibacillary
    - » Multibacillary

Ridley and Jopling classification

Based on immunologic response of the host to M leprae

Indeterminate

- TT (polar tuberculoid)
- BT (borderline tuberculoid)
- BB (borderline)
- BL (borderline lepromatous
- LL (lepromatous)

CLASSIFICATION OF LEPROSY BY WHO

- 11) Single lesion Paucibacillary HD -
  - (SLPB)

- 2) Paucibacillary HD –
  - PB

- 1 lesion, no trunk involvement, sensory loss within lesion

- 2-5 # lesions, generally asymmetrical distribution with definite loss of sensation and no more than 1 nerve trunk involvement

WHO CLASSIFICATION CONTINUED

- Multibacillary (MB) –
- More than 5 lesions with more or less symmetrical distribution, loss of sensation in various areas and may have many nerve trunks involved

HD spectrum

Classification	Zones of the HD spectrum				
Ridley – Jopling	TT	BT	BB	BL	LL
WHO Classification	PB		MB		

Indeterminate

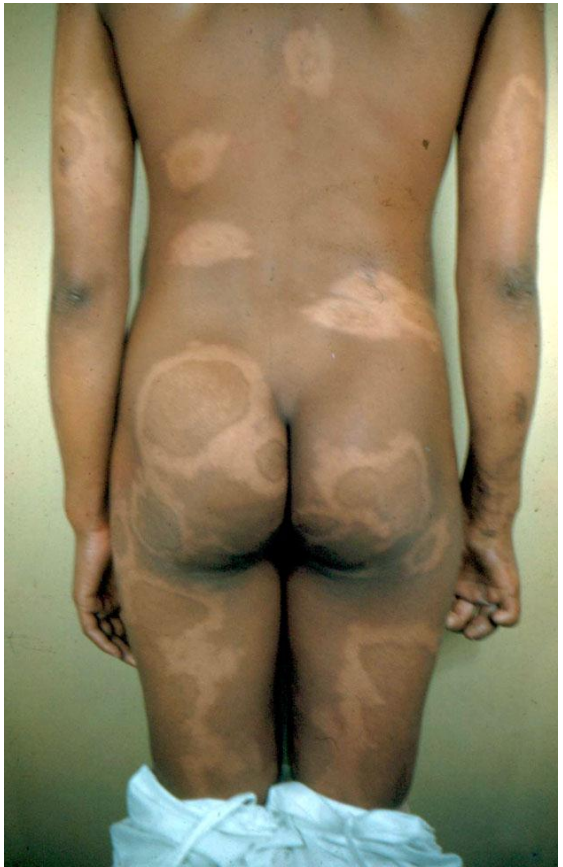
- One or few vague lesions
- Hypopigmented or erythematous macules
- Favors extremities, buttock, or face
- Minimally impaired sensation
- No peripheral nerve enlargement

TUBERCULOID
TT - Polar

- One or few lesions
- Sharply marginated, scaling macules or plaques, sometimes elevated edges
- Lesions anesthetic
- Sometimes enlarged nerve in area of lesion

BODERLINE TUBERCULOID (BT)

- Few to many erythematous, hypochromic plaques with scaling surface with well defined margins which may have small satellite lesions, usually annular



BT continued

- Lesions anesthetic asymmetrical
- Several peripheral nerves involved

BODERLINE (BB)

- Several succulent plaques with sharply demarcated central area and edges sloping into surrounding normal skin
- Large erythematous irregular infiltrated bands with central uninvolved anesthetic areas

(BB) continued

- Moderate anesthetic lesions widespread and asymmetrical peripheral nerve involvement

BODERLINE LEPROMATOUS (BL)

- Many roughly symmetrical shiny, erythematous and sometimes hypopigmented macules, papules, nodules with sloping edges.

(BL) continued

- Lesions slightly anesthetic
- Widespread and less asymmetrical peripheral nerve involvement

LEPROMATOUS LEPROSY (LL)

- Early (LL) --
  - 1) Macules small, multiple and symmetrical
  - 2) Smooth shiny surface with indistinct margins
  - 3) faintly erythematous to copper - colored

EARLY (LL) CONTINUED

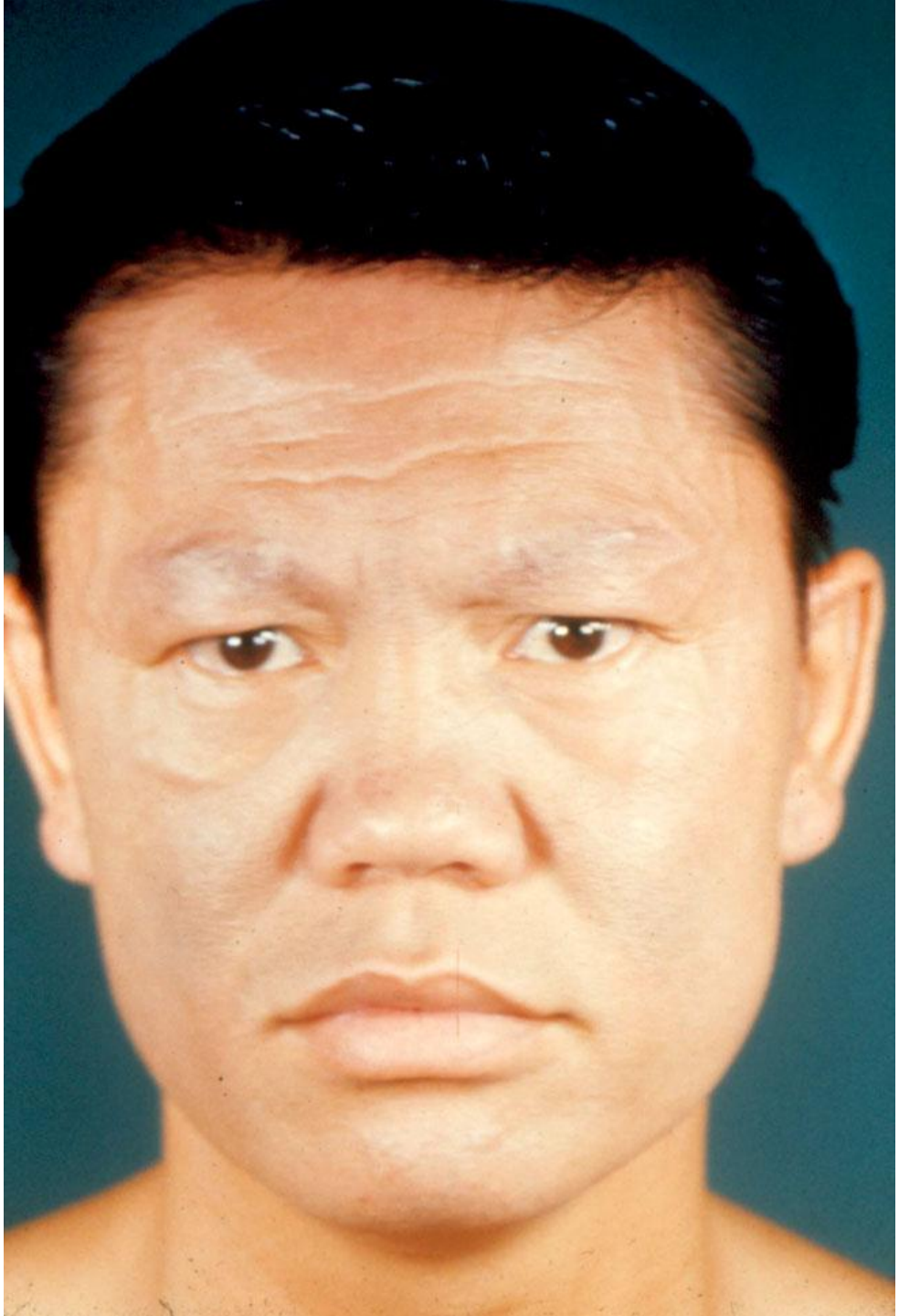
- Macules become more evident after exposure to sun
- Eyebrow hair is sparse
- Lesions hazy, indistinct and asymptomatic

INFILTRATED (LL)

- 1) Succeeds macular form
- 2) Skin is thickened, erythematous and shiny
- 3) Infiltrated plaques are glossy, soft and slope towards periphery
- 4) Sensory loss slight in infiltrated areas
- 5) Skin lesions generalized but do not occur in axilla and groin areas

(LL) continued

- Madarosis of eyebrows
- Nasal mucosal ulceration
- Usually no sensory impairment in early stages
- Later symmetrical peripheral neuropathy of arms and legs with stocking and glove anesthesia and of facial nerve
- Eye involvement (conjunctiva, cornea,and iris)



LEPROMATOUS (LL)

- Multiple and symmetrical erythematous and copper colored macules, later indurations followed by nodulation of face, especially ears and nose,extremeties,joints
- and trunk

REACTIONS IN HD

1) Reversal Reaction

BT

2) Erythema Nodosum Leprosum (ENL)

BL

LL

Reversal reaction Sx

- 1) Edema and erythema of previously visible skin lesions.
- 2) Sudden nerve damage, with swelling and pain
- 3) Fever
- 4) New, well-defined erythematous skin lesions
- 5)Edema of hands and feet

BT IN REACTION

- Allergic reaction causes edema
- Body crying to get rid of dead organisms
- Body is truly getting better

ENL SX'S

- 1) Red, tender, painful nodules
- 2) Fever, malaise
- 3) Painful neuritis, arthritis, lymphadenopathy
- 4) Eye involvement –iritis, scleritis
- 5) Edema of extremities

EYE INVOLVEMENT WITH ENL

- Infiltration of dead organisms into eye
- Nodules in iris
- Distorted pupil from reactions

LUCIO HD

- 1) Diffuse LL
- 2) Patients of Mexican and Costa Rican origin
- 3) No nodules
- 4) No wrinkles
- 5) No sweat
- 6) No body hair
  - » Bonita (pretty in Spanish)

LUCIO PHENOMENON REACTION

- Vessels become plugged causing vasculitis
- Severe ulcerations of skin

MDT adult dosages, duration, strength(s) and shortcomings

WHO MDT	Supervised monthly	Self-administered daily (4 weeks)
Pauci-abacillary (PB)	Rifampicin (600 mg)	Dapsone (100 mg) x 6 cycles in 9 months
Multi-bacillary (MB) >5 lesions	Rifampicin (600 mg) Clofazimine (300 mg)	Dapsone (100 mg) x 12 cycles in 18 months or* Clofazimine (50 mg) x 24 cycles in 36 months
ROM Single lesion PB	Rifampicin (600 mg) Ofloxacin (400 mg) Minocycline (100 mg)	Single dose

•Varies from one national leprosy control program to another. In India, where 75% of world leprosy exists, the program 12 cycles in 18 months.

US TREATMENT PLAN

- PAUCIBACILLARY

– Treat for 1 yr Rifampin 300mg – 600mg daily

Dapsone 100mg daily

Follow for 5 yrs

US TREATMENT CON'T

- Multibacillary
  - Treatment for 2 yrs -
    Rifampin 300mg – 600mg
    Dapsone 100mg daily
    Clofazimine 50mg daily

» Follow for 5 – 10 years

ADVANTAGES OF MDT

- Isolation of patient is not necessary. Disease is very difficult to transmit to another person.
- Patient is non communicable once started on MDT.
- Relapse is rare

TREATMENT OF REACTIONS

- Reversal Reaction
- Analgesics for nerve pain
- Prednisone for severe reaction

TREATMENT OF REACTIONS CONTINUED

- ENL
- Prednisone 40 – 80 mg
  - Daily single dose
  - After reaction controlled, slow tapering 5mg every 1-2 weeks

TREATMENT OF REACTIONS CONTINUED

- ENL
  - Thalidomide 50 – 200mg daily
  - Clofazimine 300mg daily for 4 – 6 wks reduced to 200 mg for several months

Clofazimine continued

- When prednisone is discontinued for 3 months then Clofazimine can be reduced to
- 50 mg daily

FOLLOW UP AFTER COMPLETION OF TREATMENT

- Paucibacillary (PB)
  - –Every six months for two years
  - –Annually for three years

FOLLOW UP AFTER COMPLETION OF TREATMENT

- Multibacillary (MB)
  - –Every six months for two years
  - –Annually for eight years

CASE STUDY

HISTORY

- 34-year-old Portuguese speaking male from Brazil
- Entered U.S. June 2005. At that time started working on farm in N.H. doing landscaping
- Spring of 2006 noted a few “mosquito bites” on his arms that itched and turned red.

HISTORY CONTINUED

- Red bumps started on arms, progressively more involved on his lower legs, trunk and face.
- Wife sent him medication from Brazil called Meticorten (prednisone), which he took for several months, with more swelling of his skin lesions.

HISTORY CONTINUED

- Therefore stopped this medication around April 2007.
- Patient saw local Dermatologist May 2007
  - Presented with diffuse large fleshy nodules on face, arms, legs, trunk, back of neck and ears.
  - Lesions on trunk were hyperpigmented, flat and diffuse

PHYSICAL EXAM

- Lesions on knees dry with light scale
- Hands diffusely swollen with lesions that are more erythematous
- Normal ear canals.
- Skin lesion extruding from right lower eyelid. – Lesion on posterior soft palate.

- WHAT COULD BE DONE TO SUPPORT

  CLINICAL DIAGNOSIS?

- Skin Biopsy
- Skin Smears
- Palpate Enlarged Nerves
- Sensory Testing

BIOPSY RESULTS

- Shave biopsy skin of left abdomen
  - Lepromatous Leprosy
  - Fite stains reveal numerous mycobacteria

FOOT AND HAND SCREENING

- Multiple foot ulcers
- Feet swollen
- No weakness in ankle or foot
- No loss of protective sensation
- Hands residual texture with sensory testing
- No muscle weakness
- No tender nerves

TREATMENT PLAN

- Clofazimine 100mg daily
- Rifampin 300 mg daily
- Ofloxacin 400mg daily
- Prednisone 20 mg daily

REFERRALS

- Infectious Disease
- Otolaryngology
- Ophthalmology

FURTHER DIAGNOSTIC TESTS

- G6PD
- CBC
- LFT’s
- Eosinophilia
- Hep B Surg AB
- Strongylidiasis AB
- Stools O & P
- Crypto/Giardia
- PPD
- CXR
- Cat Scan/Thorax
- Toxocara Canis AB

Foot drop & ulnar damage

11/85

PATIENT PROFILE Ins M'C

NAME:

M.R.N.# 824973 -3 H.D.-I.D.# 09033010

SS# 022-66-6392

TELEPHONE 561-3158 AGE 04/02/53 32yr.

1/96 508-587-1396

ADDRESS ~~255 MARION ST~~

~~APT #2~~

E. BOSTON, MA 02128

~~6/92 19 Walnut Place #205~~

~~Revere, Ma. 02151~~

~~12/93 22 Shirley Ave Revere Ma~~

~~10/95 9 Rogers Ave #2~~

~~Lynn, Ma 01902-3802~~

1/96 19 Orchard Ave, Brockton 02401

COUNTRY OF ORIGIN CAMB.

NCP

DX: (TL) BT BB BL LL

2/85

DATE

1967?

BX RESULTS: 11/30/84 - Neg

2/22/85

1/29/86 (BMAHC) - a perivascular lymphohistiocytic infiltrate is present in upper dermis as well as occ. granulomas. No AF organism. findings are consistent c̄ T-Type leprosy

Surg 1/14/88 Amp (R) index finger at PIP

1/4/89 SKIN Smears

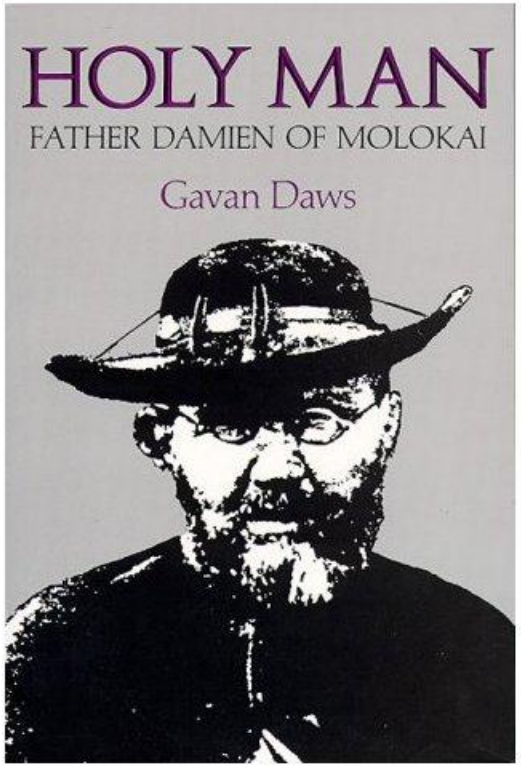
Eye Exam: 1/7/86

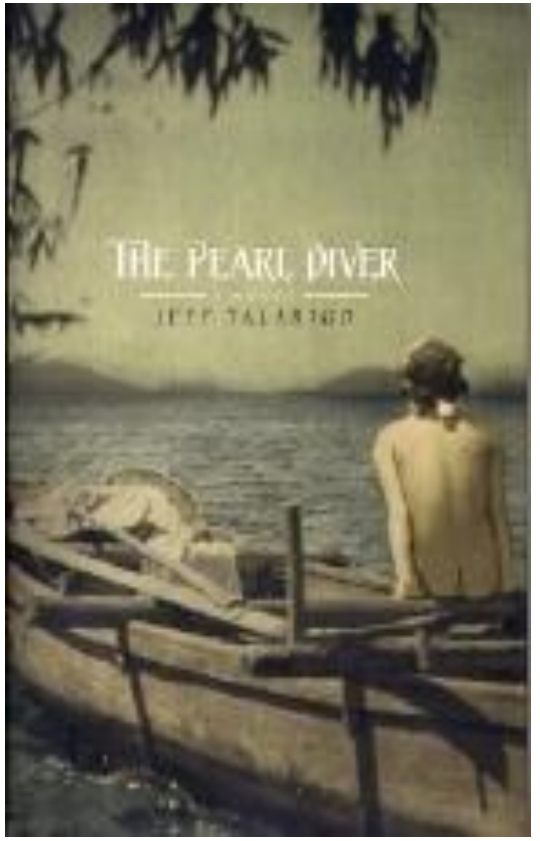
FNC: [illegible] 3/[illegible]/89

IMPORTANT LESSON TO REMEMBER

- Leprosy can be cured
- Leprosy is a slow disease
- Leprosy takes a long time to come and a long time to go away
- Medication can cure leprosy patches, but not deformity

SUGGESTED READING





CHINESE PROVERB

- “What we hear, we forget,
- What we see, we remember,
- What we do, we know.”