The History of Leprosy in Hawaii
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Content Attestation
I, Vernon Ansdell, M.D., hereby declare that the content for this activity, including any presentation of therapeutic options, is well balanced, unbiased, and to the extent possible, evidence-based.
Leprosy: History

- Recognized in ancient civilizations of China, Egypt, and India
- First written record 600 BC Indian writings
- Feared and misunderstood throughout history
  - Incurable, disfigurement, disability, isolation, psychological trauma, poverty

Leprosy: History

- Many references to “lepers” in the Bible
- “Curse”, “punishment from the Gods”, hereditary disease, sexual perversion
- Middle Ages (10th – 15th centuries): Inflicted forced to wear special clothing and ring bells to warn others “unclean”

Leprosy: History

- As many as 1 in 30 infected in some areas
- Rapid decline in the 15th century in Europe
  - Famine of 1315?
  - Black Death 1347-1349?
  - Not due to loss of virulence of M leprae
**Leprosy: History**

- **Diagnosis:**
  - M. leprae identified by Hansen in Norway in 1873 “Hansen’s Disease”

- **Treatment:**
  - Chaulmoogra oil up to 1941
  - Sulfones 1940’s (Dapsone)
  - Multi drug treatment (MDT) 1970’s

**Leprosy: Microbiology**

- Mycobacterium leprae (M leprae)
- Acid fast bacillus
- Multiplies slowly
- Obligate intracellular organism
- Cannot be cultured in artificial medium
- Grows best at 27°C - 33°C cooler areas – skin upper respiratory mucous membranes, cutaneous nerves

- Armadillos have core body temperatures of 34°C. Enzootic in wild armadillos in south central USA

- Found in chimpanzees, cynomolgus, macaque monkey, sooty mangabey monkeys
Leprosy: Transmission

- Respiratory route
- Large numbers of bacilli in nasal secretions from multibacillary disease
- Occasional transmission via broken skin
- Contact with armadillos (handling, killing, eating in some cases)

Leprosy: Classification and Terminology

- Ridley-Jopling Classification
  - Tuberculoid (TT)
  - Borderline tuberculoid (BT)
  - Borderline (BB)
  - Borderline lepromatous (BL)
  - Lepromatous (LL)
  - Indeterminate (I)

Tuberculoid Leprosy (TT)

- 1–2 large macular hypopigmented or erythematous anesthetic lesions
- Well defined, often raised margins
- Paucibacillary
- Neural involvement common
Borderline Tuberculoid (BT)

- Sharply defined macules
- More numerous than TT
- Paucibacillary
- Anesthesia less than (TT)

Borderline Borderline (BB)

- Numerous erythematous, irregular plaques. Some “punched out” lesions
- Moderate anesthesia
- Multibacillary

Borderline Lepromatous (BL)

- Numerous erythematous, irregular macules, papules, plaques and nodules
- Often symmetrical
- Anesthesia often absent
- Multibacillary
Lepromatous Leprosy (LL)

- Diffuse infiltration and thickening of the skin
- Loss of eyebrows and eyelashes
- Nodular thickening of the earlobes
- Nasal stuffiness
- Septal perforation and collapse (Saddle nose)
- Hypogonadism (Testicular involvement)
- Multi bacillary

Leprosy: Peripheral Nerve Involvement

- Ulnar nerve at the elbow
- Median and superficial radial nerve at the wrist
- Great auricular nerve at the neck
- Common peroneal nerve at the popliteal fossa

Leprosy: Eye Involvement

- Lagophthalmos
- Drying of the cornea
- Corneal abrasion
- Corneal ulceration
- Nerve damage to eyelid muscles and cornea
Leprosy: Late Clinical Findings
- Claw hand, foot drop, facial paralysis, lack of eyebrows and eye lashes, collapsed nose, perforated nasal septum

Leprosy: Clinical Clues
- Hypopigmented or reddish skin patches
- Diminished sensation within skin patch
- Tingling, numbness in hands and feet
- Painless wounds and burns on hands and feet
- Tender, enlarged peripheral nerves
- Swelling of earlobes and face

Leprosy: Laboratory Diagnosis
- Skin biopsy – culture to exclude tuberculosis and non-tuberculosis mycobacteria
- Polymerase chain reaction (PCR)
- Serologic tests (PGL-1 antibody) not generally available
Risk Factors for Leprosy

- Close, prolonged contact
- Age: Bimodal – increased risk between 5 and 15 years; risk increasing again after age 30
- Genetic influences
- Armadillo exposure
- Type of leprosy in index patient

New Cases of Leprosy – Reported to WHO January 2012

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<tr>
<th>Region</th>
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<td>South East Asia</td>
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<td>India</td>
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<td>South Sudan</td>
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</table>
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Leprosy in USA
- 205 new cases in 2010
- 75% in immigrants
- Some cases acquired by travelers abroad
- Some cases from exposure to infected armadillos
- Often there is no history of exposure

Hawaii: Late 1700’s
- Population of Hawaii: 250,000 – 300,000
- Era of European contact.
  - Arrival of European explorers – Captain James Cook

Hawaii: Late 1700’s
- Rapid depopulation
- Famine, battlefield deaths, infectious diseases
- Plague, smallpox, influenza, measles, mumps, cholera, pertussis, gonorrhea, syphilis, tuberculosis

Hawaii: Late 1700's
Population fell from 250,00 – 300,000 to 31,000 by 1896

Leprosy in Hawaii
- Introduced by Chinese plantation workers?
  Mai Pake (Chinese sickness)
  Mai Ali'i (Chief's sickness)
- First described in Hawaii as early as 1823
- Well established by 1830's and a major concern by 1860's
  "epidemic proportions"

Leprosy in Hawaii
- Marked prevalence in native Hawaiians
  "Vanishing population" 50,000 – 60,000
- King Kamehameha V signed into law
  "Act to Prevent the Spread of Leprosy"
  3 January 1865
- Set apart land for the isolation and segregation of patients with leprosy
- Kalihi Hospital and Detention Station
Kalaupapa

- Isolated peninsula on the north shore of the island of Molokai
- Bordered by 2,000 feet cliffs and open ocean “natural prison”
  - Fertile soil suitable for taro, sweet potatoes, fruits, and livestock
  - Fish from the ocean
  - Freshwater from Waikolu Valley

Problem was that, because of their disease, most of the patients were physically unable to farm or fish!!
Leprosy in Hawaii

- January 6, 1866 – 9 men and 3 women with leprosy were forcibly exiled to Kalawao
  - Very limited shelter
  - No tools or materials
  - No medicines
  - No medical providers
  - No authority
- By October 1866, 101 men and 41 women had been exiled to Molokai

Leprosy in Hawaii

- Patients hid to avoid capture and transportation to Kalawao and Kalaupapa (promoted disease spread)
- Reports of a state of lawlessness – gambling, debauchery, alcohol abuse
  “Aole kanawai ma keia wahi.”
  - In this place there is no law.

Leprosy in Hawaii

- Some attempts by Board of Health to improve the situation.
  - Built schools
  - Dispensary
  - Cook house
  - Morgue
  - Jail
  - Churches
  - Supported na koku (helpers)
Father Damien

- Born Joseph de Veuster in Belgium in 1840
- Came to Hawaii as a missionary in 1864
- Worked on the Big Island to 1873
- Arrived in Kalaupapa May 1873

Father Damien

- Arrived on Molokai 10, May 1973
- Worked to restore order and dignity and provide effective medical care
- Learned to speak Hawaiian
- Helped to build churches, houses, schools and hospital
- Made coffins, dug graves
- Destroyed distilleries

Kalaupapa


Father Damien

- Physical and spiritual needs
- 1884 realized that he had leprosy
- Died April 15, 1889
- Declared a Saint in 2009

Sister Marianne Cope

- Born in Germany
- 1883: Came to Hawaii to care for patients with leprosy (Oahu and Maui)
- 1888: Moved to Kalaupapa to care for Father Damien and take over his role
- 1918: Died of natural causes
- 2012: Declared a Saint
Leprosy: Early Treatment in Hawaii

- 1879 Hoang Nan Pills
  - Treatment required abstinence from poi, raw fish, and kawa!
- 1882 Goto baths (hot baths containing herbs, oral herbs)
- Early 1900’s – Chaulmoogra oil
  - Oil from seeds of Hydnocarpus trees
  - Described in ancient Hindu and Chinese literature
  - Documented antibacterial properties

Chaulmoogra Oil

- Oral, subcutaneous, intramuscular routes
- Poorly tolerated
- Variable efficacy
- Hundreds of patients “cured” and released from Kalihi Hospital and Kalaupapa

Chaulmoogra Oil: Joseph Rock

- Botanist at the University of Hawaii
- Searched for the Chaulmoogra tree in Burma, Thailand and Assam
- Brought back seeds to Hawaii
- Thousands of Chaulmoogra trees planted on Windward Oahu
Chaulmoogra Oil: Alice Ball

- Postgraduate research at the University of Hawaii
- Chemist who developed a process to isolate the ethyl esters of the fatty acids in chaulmoogra oil
- Injectable chaulmoogra extract was the preferred treatment for leprosy until the introduction of sulfones in 1940s
- First woman and first African American to graduate from the University of Hawaii with a master’s degree

Kalaupapa/Kalawao

- Total residents more than 8,000
- Peak population was 1,100 in 1890
- By 1900, the number of new leprosy patients in Hawaii was declining
- Total population was 888 in 1903
  - 541 males, 347 females
  - 459 Hawaiian men, 338 Hawaiian women
- Confinement laws lifted in 1969
- Currently 17 patients still living in Kalaupapa (by choice)