Neurosyphilis & the Clinical Motor Speech & Cognitive-Linguistic Deficits & Treatment

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What is Syphilis?

- From the Greek word “syphlos”, meaning crippled or maimed
- A sexually transmitted disease caused by the bacteria *Treponema palladium*
- Referred to as the “Great Imitator”
- **Prevalence:** In the U.S. alone, health officials reported over 36,000 cases in 2006
  - Primarily occurs in individuals 20-39 years of age

http://www.merck.com/mmpe/sec14/ch194/ch194i.html
What is Neurosyphilis?

- An infection of the brain or spinal cord
- Occurs in individuals who have untreated syphilis
- Caused by the bacteria *Treponema pallidum*, which causes syphilis
- Develops about 10 - 20 years after a person is first infected with syphilis
- Not everyone who has syphilis will develop this complication (6.5% of untreated syphilis cases lead to neurosyphilis).

http://www.nlm.nih.gov/medlineplus/ency/article/000703.htm#visualContent
Symptoms of Neurosyphilis

- Abnormal gait
- Blindness
- Confusion
- Dementia
- Depression
- Headache
- Incontinence
- Inability to walk
- Irritability
- Loss of muscle function
- Mental decline
- Paralysis
- Poor concentration
- Seizures
- Stiff neck
- Tremors
- Visual disturbances
- Weakness/numbness of the lower extremities

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Diagnostic Tests & Exams

- Confirmatory Signs include:
  - Muscle atrophy
  - Muscle contractions
  - Abnormal reflexes
  - Blood tests to detect substances produced by the bacteria that cause syphilis

- Other tests include:
  - Fluorescent treponemal antibody absorption (FTA-ABS)
  - Rapid plasma reagin (RPR)
  - Treponemapallidum particle agglutination assay (TPPA)

* it is important to test the spinal fluid for signs of syphilis
Tests to look for problems with the nervous system include...

- Cerebral angiogram (uses dye & x-rays to examine how blood flows through the brain)
- Head CT scan
- Lumbar puncture ("Spinal tap")
- MRI scan of brain, brainstem, or spinal cord
4 forms of Neurosyphilis

- Asymptomatic
- General Paresis (Parenchymatous or dementia paralytica)
- Meningovascular
- TabesDorsalis (Locomotor ataxia)
Cognitive-Linguistic & Motor Speech Symptoms of General Paresis Neurosyphilis

- Results when chronic meningoencephalitis causes destruction of cortical parenchyma
- Develops 15 to 20 yr after initial infection and typically does not affect patients before they are 40-50 years old
- Behavior progressively deteriorates, sometimes resembles a mental disorder or dementia
- Irritability, difficulty concentrating, deterioration of memory, defective judgment, headaches, insomnia, fatigue, and lethargy are common
- Seizures, aphasia, and transient hemiparesis are possible. Patients may become emotionally unstable and depressed and have delusions
- Tremors of the mouth, tongue, hands, and whole body may occur; other signs include pupillary abnormalities, dysarthria, hyperreflexia, and, in some patients, extensor plantar responses
Cognitive-Linguistic & Motor Speech Symptoms of Tabes dorsalis (locomotor ataxia)

- Involves slow, progressive degeneration of the posterior columns and nerve roots
- Develops 20 to 30 yr after initial infection
- Usually the earliest symptom is an intense, pain in the back and legs that occurs irregularly
- Gait ataxia, hyperesthesia, and paresthesia may produce a sensation of walking on foam rubber
- Most patients with tabes dorsalis are thin and have characteristic sad facies and Argyll Robertson pupils (pupils that accommodate for near vision but do not respond to light)
- Optic atrophy may occur
- Hypotonia, hyporeflexia, impaired vibratory and joint position sense, ataxia in the heel-shin test, absence of deep pain sensation, and Romberg's sign
- Tabes dorsalis tends to be intractable even with treatment
Medical Treatment of Neurosyphilis:

- Penicillin
- Alternative = parenteral ceftriaxone
- Given in various ways:
  - Injected
  - Probenecid (a drug that increases uric acid)

- Follow-up
  - blood tests and lumbar punctures for CSF fluid analysis at 3, 6, 12, and 24 months to make sure the infection is gone

Prognosis
- Treatable if detected early
- However, it is still considered a life-threatening complication of syphilis
- The success of treatment is dependent on the severity level of the neurosyphilis before treatment is given

Shann & Wilson, 2003
Motor Speech Symptomatology in patients with Neurosyphilis:

- Possible cranial nerves (V, XII) involvement
- Flaccid & Ataxic (28%)
- Dysarthrias, and progressive supranuclear palsy
- Other neurological problems that affect speech and voice

(Duffy, 2005)
Management of Flaccid Dysarthria in Patients with Neurosyphilis:

- Due to muscle weakness, unique treatments tend to focus on:
  - Increasing strength
  - Compensation for muscle weakness
  - Postural adjustments and/or prosthetic aids may be used in treatment

- Treatments should be aimed at the following components of speech:
  - respiratory
  - phonatory
  - Resonatory
  - articulatory

(Duffy, 2005)
Management of Flaccid Dysarthria in Patients with Neurosyphilis Continued:

- **Symptom:** possible reduced loudness
  - Treatment for increasing respiratory function includes:
    - Deep inhalation
    - Controlled exhalation
    - Inspiratory monitoring
    - Increased force

- **Treatment for closing the vocal folds**
  - Possible surgery (i.e. arytenoid adduction surgery)
  - Voice amplifiers (temporary or permanent)
  - Palatal lift treatment
  - Pharyngeal flap surgery

(Duffy, 2005)
Case Study:

- 61 year old man, served abroad in the armed forces
- Diagnosis of symptomatic neurosyphilis (Sept. 1997)
- Diagnosed with expressive dysphagia and a left sided hemiparesis

**Symptoms:**
- “Out of character” behavior
- Impaired use of right hand
- Marked short-term memory loss
- CSF spaces disproportionate to patient’s age
- Frequent falls and delusions

**Treatment:**
- Allergic to penicillin; treated with ceftriaxone. Received 1 gram 1x/day for 14 days

(Shann, 2003)
Neurosyphilis Effects on Vision:

- Ophthalmic: Blurred vision, reduced color perception, impaired acuity, visual dimming & photophobia 17%
- Pupillary changes: Argyll Robertson pupils 43%
- Optic atrophy 7%
  (Timmermans, 2004)

- A 44 year old man with neurosyphilis reported that his vision was “like looking through tissue paper” in his left eye
  (Smith et al., 2006, p. 36)

- Neurosyphilis: Pupil
References

- [http://www.nlm.nih.gov/medlineplus/ency/article/000703.htm#visualContent](http://www.nlm.nih.gov/medlineplus/ency/article/000703.htm#visualContent)