# NURS 821 Neurological Disorders

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Lecture 11

Part 5 Neurodegenerative Diseases

## Amyotrophic Lateral Sclerosis (ALS)

- Definition: a progressive degenerative disorder of motor neurons in the spinal cord, brainstem, and motor cortex.
- Etiology: may be caused by a viral-immune process.
- Incidence: a common disease; annual incidence rate of 0.4 to 1.76 per 100,000 population
- M>F
- Age of onset is > 50 years

## Amyotrophic Lateral Sclerosis – Incidence Continued…

- Incidence increases with each decade of life
- In about 5% of cases the disease is familial (autosomal dominant)
- Occurs in a random pattern throughout the world except for a dramatic clustering of patients among inhabitants of the Kii peninsula in Japan and in Guam.

Source: Victor & Ropper (2001)

## Amyotrophic Lateral Sclerosis- Pathophysiology

- Principal finding- loss of nerve cells in the anterior horns of the spinal cord and motor nuclei of the lower brainstem
- Lost cells are replaced by fibrous astrocytes
- Many surviving nerve cells are small, shrunken, and filled with lipofuscin
- Mutations in gene SOD1 have been found in familial cases

## Amyotrophic Lateral Sclerosis

- Early clinical features:
  - Stiffness of fingers, and slight weakness or wasting of hand muscles are the first signs
  - Awkwardness in tasks requiring fine finger movements (e.g. buttons and ignition keys)
  - Cramping of muscles, beyond normal
  - Fasciculations of the muscles of forearm, upper arm, and shoulder girdle

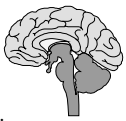
## Amyotrophic Lateral Sclerosis

- Early clinical features cont'd…
  - In weeks to months, the symptoms affect both sides of the body
  - Soon the diagnostic triad of atrophic weakness of the hands and forearms, slight spasticity of the arms and legs, and generalized hyperreflexia become evident – all in the absence of sensory change
  - At this time diagnosis is inevitable

## Amyotrophic Lateral Sclerosis

- Later clinical features:
  - Muscle strength and bulk diminish
  - Tendon reflexes become hyperreflexic
  - Atrophic weakness spreads to the neck tongue, pharyngeal, and laryngeal muscles
  - Eventually, the trunk and lower extremities are involved

## Parkinson's Disease



- Definition: Group of insidious chronic, progressive conditions called motor system disorders
- Etiology: Death of dopamine-producing neurons; dopamine depletion by drugs, e.g. methyldopa Related to vascular, viral, and metabolic factors
- Consequence: Neurons fire out of control
- Manifestations: inability to control movement: tremors; limb and trunk rigidity; bradykinesia; postural instability; difficulty with walking, talking, simple task performance (NINDS, 2000)

## PD - Incidence

- Common degenerative disease
- In North America, approximately one million persons are afflicted with Parkinson Disease
- Approximately 1% of the population over the age of 65 years is affected
- Typically begins between age 40 and 70, infrequently before age 30
- Peak onset is at age 60

## PD - Incidence

- M>F
- Whites:Blacks 4:1
- Whites:Asians 3:1
- Familial cases have been documented, but are considered rare
- More frequently found in industrialized countries

Source: Victor & Ropper (2001)

## PD - Etiology

- Epidemiological data suggest vascular, viral, and metabolic factors
- Related to loss of dopamine-producing brain cells
- It was observed in human opiate addicts and in monkeys, that a neurotoxin (known as MPTP) can produce irreversible signs of PD.

## Parkinson's Disease Pathophysiology

- Microscopic findings include:
  - Protein deposits in the CNS
  - Mutations in the gene asynoclein (autosomal dominant) found in patients with familial PD
  - Loss of pigmented cells in the substantia nigra (by approximately 66%)
  - Remaining pigmented cells contain Lewy bodies

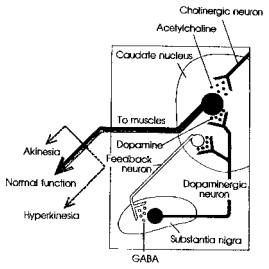
## Parkinson's Disease Manifestations

PD results from:

- Degeneration of the dopaminergic nigrostriatal pathway ⩾ causing dopamine depletion in the basal ganglia ⩾ resulting in relative excess cholinergic activity in the feedback circuit ⩾ hypertonia (tremor and rigidity) and akinesia

## Parkinson's Disease Pathology

Feedback circuit:




## Parkinson's Disease Manifestations

- Very early clues:
  - Reduced blink rate. (Usual blink rate is 12 – 20/min. PD rate is 5-10 blinks/minute.)
  - Lack of arm swing (Myerson glabellar sign)
  - Digital impedance (slow rapid alternating movements)
  - Resting tremor

## Parkinson's Disease Manifestations

- Expressionless face (due to reduction in movement of small facial muscles)
- Bradykinesia, or slowness of movement
- Postural instability or impaired balance and coordination
- One side of the body is typically involved (usually left side) before the other

## Parkinson's Disease

- May have difficulty:
  - Completing simple tasks (buttoning of shirt)
  - Walking (shuffled gait)
  - Talking (soft, monotone)