Acute and Chronic Neurological Diseases: Quick Notes

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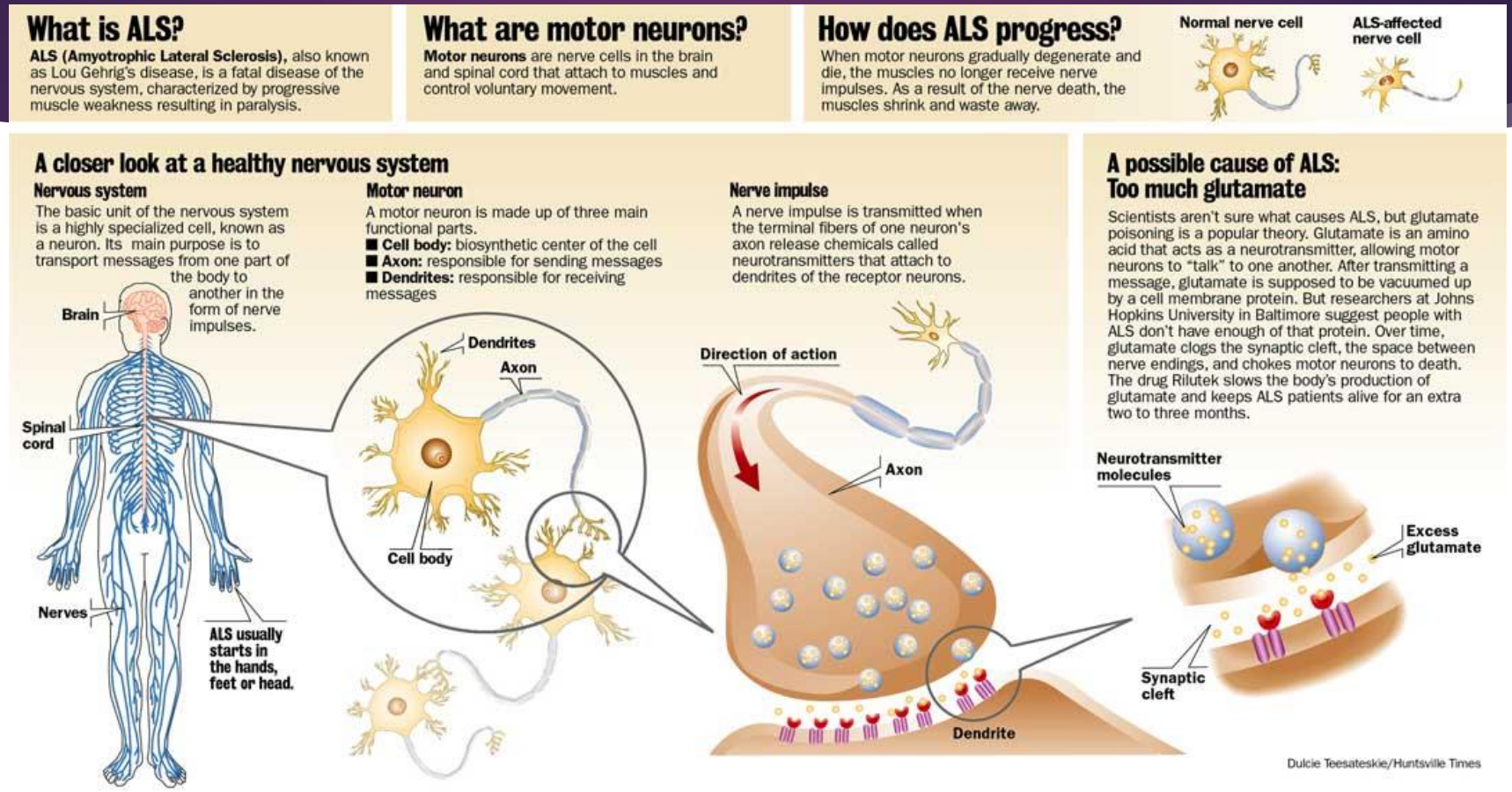
Neurological Conditions

- Amyotrophic Lateral Sclerosis

Amyotrophic Lateral Sclerosis



Amyotrophic Lateral Sclerosis

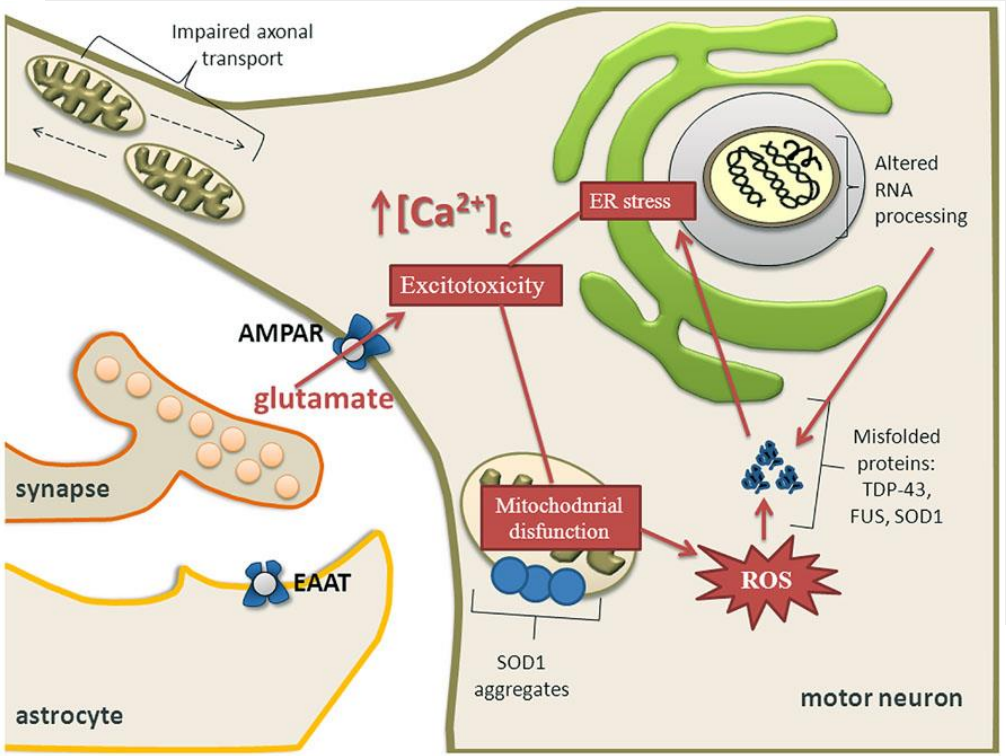
- Amyotrophic lateral sclerosis (ALS) is the most common motor neuron disease in adults
- The incidence of ALS is approximately **1–2.6 cases per 100 000 persons annually**, whereas the prevalence is approximately **6 cases per 100 000**.
- Those who were in the Gulf War are more likely to get ALS compared with other veterans.
- 20% more common in men than in women
- Typically midlife, between 40-70 years- most common Caucasian
- Life Expectancy is 2-5 years, more than ½ live longer than 3 years

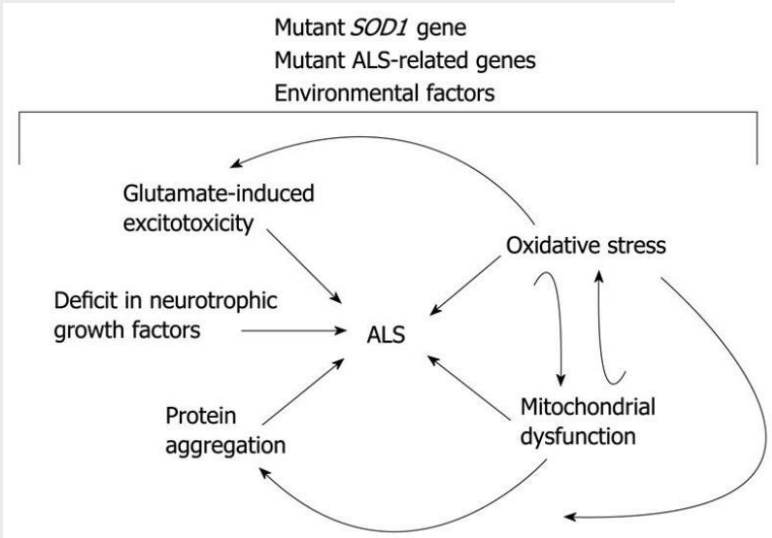
On June 1, 2021, a team of scientists let by the NIH and the Uniformed Services University announced it had discovered a unique form of genetic ALS that affects children as early as age 4 years. This childhood form of ALS is linked to the gene ***SPTLC1***

Etiology

- Sporadic , unkown cause
- Genetic in 5-10% of cases-

an inherited autosomal trait





Etiology

- Theoretical Models
  - Excitotoxicity- Glutamate defect in possibly metabolism, transport or storage
  - Oxidative stress caused by free radicals
  - Autoimmune-antibodies to calcium channels, activated T lymphocytes , Monoclonal paraproteinemia
  - Cytoskeletal defects or abnormalities
  - Neurofilament abnormalities- abnormal accumulation and damage to the structure

Rehabilitation Goals

- Prevent complications of immobility
- Strengthen unaffected muscles
- Saliva management
- Dysphasia
- Aspiration prevention, Assisted Cough
- Communication, plan early before need for ventilator
- Adaptive techniques and equipment
- Depression, coping with progressive loss
- Preparation for disease progression
- Preparation for death

Focus on Palliative Care

- Key Decisions must be discussed in advance of crisis
  - G-Tube
  - Mechanical Ventilation
  - Advance Directives
- Teaching Plan for Patient and Caregiver/Family
  - Symptom Management
  - Reduce Complications
- Assistive Devices to maintain independence
- Referrals for support groups and other resources

Nursing Process

- Assessment
  - Full Health History
  - Family Incidence of ALS
  - Onset of Symptoms and Progression
- Current Function & ADLs
  - Gait, strength and stability
  - Flaccidity, spasticity
  - Eval Swallowing and chewing
  - Eval Respiratory Status
  - Bowel and Bladder function
  - Skin Assessment
  - Observe Family Interactions

Plan of Care

- **Nursing Diagnoses**
  - Impaired physical mobility re: muscle wasting, weakness and spasticity
  - Self Care deficit
  - Impaired Communication re: impairment of muscles for speech
  - Ineffective breathing pattern re: impairment of diaphragm and accessory muscles

Plan of Care

- **Nursing Diagnoses**
  - Altered nutrition: Less than required re: bulbar muscles
  - Potential for anxiety
  - Risk for ineffective coping
  - Interrupted family processes re: change in health of family member, modifications of family roles and foreseen loss of family member
  - Risk for caregiver strain

Interventions

- Maintain Independence
- Limit complications from progressive loss of function
  - Ineffective breathing
  - Impaired swallowing and decrease in nutritional intake
  - Impaired communication
  - Maintain skin integrity
  - Patient and Family Education and support in decision making
  - Pain management

Treatment for ALS

- **The U.S. Food and Drug Administration (FDA) has approved drugs to treat ALS:**

- **Riluzole** (Rilutek) is an oral medication believed to reduce damage to motor neurons by decreasing levels of glutamate, which transports messages between nerve cells and motor neurons. Clinical trials in people with ALS showed that riluzole prolongs survival by a few months, particularly in the bulbar form of the disease. Individuals with swallowing difficulties may prefer the thickened liquid form (Tiglutik) or the tablet (Exservan) that dissolves on the tongue.
- **Edaravone** (Radicava) is given by intravenous infusion and has been shown to slow the decline in clinical assessment of daily functioning in persons with ALS.

medications to help manage symptoms of ALS, including muscle cramps, stiffness, excess saliva and phlegm, and the pseudobulbar affect (involuntary or uncontrollable episodes of crying and/or laughing, or other emotional displays). Drugs also are available to help individuals with pain, depression, sleep disturbances, and constipation.

Treatment for ALS

- Medications can be prescribed to help manage symptoms of ALS:
  - Muscle cramps
  - Stiffness
  - Excess saliva and phlegm
  - Pseudobulbar affect (involuntary or uncontrollable episodes of crying and/or laughing, or other emotional displays).

- Drugs also are available to help individuals with:
  - Pain
  - Depression
  - Sleep disturbances
  - Constipation

Non-Pharmacological Treatment

- **Communications support**
  - As ALS progresses, speech therapists can help people maintain the ability to communicate.
  - Devices such as **computer-based speech synthesizers** use eye-tracking technology and can help people develop ways for responding to yes-or-no questions with their eyes or by other nonverbal means
  - A **brain-computer interface (BCI)** is a system that allows individuals with ALS to communicate or control equipment such as a wheelchair using only brain activity.

- **Nutritional support**
  - Small meals that provide enough calories, fiber, and fluid
  - Avoid foods that are difficult to swallow
  - Suction devices
  - Feeding tube

- **Breathing support**
  - Noninvasive ventilation (NIV) refers to breathing
  - Mechanical ventilation (respirators)

- **Rehabilitation Team**
  - Physical Therapy
  - Occupational therapy
  - Speech Therapy