Huntington's Disease Assessment and Interventions

Huntington's disease is a degenerative neurologic disorder that affects both men and women. This autosomal dominant disease has a late onset between 30 to 50 years of age. The deficiency of acetylcholine and GABA, an inhibitory neurotransmitter, causes choreiform movements and cognitive decline. As the disease progresses, gait deteriorates and decrease muscle control increases the risk of aspiration. Interventions for Huntington's disease include genetic counseling in addition to the administration of neuroleptics and tetrabenazine. Nursing considerations include educating the patient about the stages of Huntington's disease and recommending a high calorie diet to prevent malnutrition.



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Assessment

Decreased GABA

Down-arrow GABA-goose

Huntington's disease is associated with decreased serotonin and GABA. Decreased levels of serotonin affect memory and learning abilities. Decreased GABA causes increased dopamine and presents as chorea, which is defined by excessive and abnormal involuntary movements. The disease may also increase levels of glutamate, an excitatory neurotransmitter and contribute to excessive motor movements.

Appears around Age 40

(40) Ounce

Symptoms of Huntington's disease normally appear around age 40. If the patient is symptomatic, genetic testing may be done to confirm the disease. Aspiration pneumonia and heart disease are the leading causes of death, which usually occurs 10 to 20 years after onset of symptoms.

Slow Cognitive Decline

Snail Cogs Decline

This progressive, degenerative brain disorder may cause slow cognitive decline and eventually develop into dementia. Decreased levels of serotonin may lead to memory and learning impairments. The patient's perception, memory, attention span, and ability to learn deteriorates as the disease progresses. The abilities to eat and talk decreases and creates frustration for the patient.

Choreiform Movements

Korean-flag

The increased levels of dopamine cause choreiform movements in the face, limbs, and body that are difficult to control. As the disease progresses, the movements worsen and ambulation gradually becomes impossible.

Non Repetitive Jerks

Nun Repeating Jerks

Increased levels of dopamine result in the non repetitive jerks seen in patients with Huntington's disease. These movements are characterized as odd, uncontrollable, and excessive. The amount of non repetitive jerking movements increases as the disease progresses.

Interventions

Neuroleptics

Nerve-leopard

Neuroleptics, such as haloperidol and risperidone, may be used to decrease the symptoms of Huntington's disease by blocking dopamine receptors and decreasing choreiform movements. However, these medications do not slow down disease progression.

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Tetrabenazine

Tetris-bonsai-tree

Excessive levels of dopamine cause chorea in patients with Huntington's disease. Tetrabenazine decreases dopamine transmission in the brain to significantly moderate chorea.

Genetic Counseling

Gene Counselor

Since Huntington's disease is an autosomal dominant disorder, genetic counseling is recommended to confirm the presence of disease. A positive test indicates that a person will develop the disease but the timing and extent is unknown. DNA testing may be done on fetal cells retrieved by amniocentesis or chorionic villus sample. Since there is no cure for Huntington's disease, genetic counseling of expectant families is the only method of decreasing prevalence of the disease.

Considerations

Stages of Disease

Stages of Hunters

Huntington's disease is a progressive condition that is classified in stages. Each of the three stages lasts about 5 years. During stage 1, the patient may present with psychological and neurological deficits but is able to maintain independence. During stage 2, the patient may experience increased choreiform movements and will require assistance with ADLs. The majority of patients in stage 3 of Huntington's disease are completely dependent on others to perform ADLs.

High Calorie Diet

High Pancakes

Huntington's disease causes choreic movements that increase the patient's caloric requirements. In order to maintain body weight, the patient may need up to 5000 calories/day. As the disease progresses, maintaining nutritional requirements may necessitate a switch to pureed foods or thickened liquid diets since patients may have increased difficulty swallowing.

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