

Huntington's chorea or disease is an inherited progressive neurological disorder with no cure that ultimately results in death. The average life span after diagnosis is seventeen years. The disease is characterized by progressive dementia and abnormal jerky physical movements referred to as chorea. It can occur in childhood, but most commonly appears in the third, fourth, or fifth decade of life.

The root of the disease lies primarily in the caudate nucleus. There is a defective repetitive sequence on the Huntingtin gene (yes, this is spelled correctly!) This altered gene results in an altered form of the Huntingtin protein. It is this mutant protein that damages the brain's neurons.

These patients may be prone to aspiration due to involvement of the pharyngeal muscles. In fact, the most significant motor symptom may be dysphagia which can result in malnourishment. Succinylcholine should be used judiciously in this population because of reports of decreased plasma cholinesterase and resulting increased sensitivity to the drug. Nondepolarizing muscle relaxants should also be carefully titrated due to the potential for *extreme* sensitivity. Prepare the patient and family for potential postoperative ventilation.