

Progressive,
increasing dementia
and chorea (jerking,
rapid, involuntary



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Progressive, degenerative, inherited neurologic disease characterized by increasing dementia and chorea (jerking, rapid, involuntary movements) and speech problems. Decrease in acetylcholine, resulting in relative excess of dopamine. Excess dopamine causes the excessive uncontrolled movements. Onset at about 40 yrs of age. If there are more genetic mutations found on the affected chromosome, clinical manifestations can occur even earlier. Autosomal Dominant. 50% chance of passing. Huntingtin (HTT) found on Chromosome 4. Genetic testing looks at the amount of times "CAG" is repeated in the coding for glutamine. In Huntington's "CAG" is repeated over 36 times when normally, it is only repeated 10-35 times. Magnetic resonance imaging (MRI) or Computerized tomography (CT) used to identify specific areas of the brain effected by the disease. Used to rule out other conditions. Psychiatric Evaluation. Emotional State. Coping Skills. Substance abuse? Movement/Psychiatric/Sensory. Movement. Unsteady Gait and involuntary movements. Slurred speech. Difficulty swallowing. Chorea- purposeless, dance-like. Psychiatric. Personality changes. Mood swings. Depression. Forgetfulness and Impaired judgment. Sensory. Hearing. Eye Movement. Significant weight loss. Huntington's Disease cannot be cured. Treatment addresses disease manifestations.

Movement Disorders: Tetrabenazine (Xenazine) is approved by the FDA to suppress the involuntary jerking and writhing movements. A serious side effect is the risk of worsening or triggering depression.

Psychiatric Disorders: Antipsychotics, such as haloperidol (Haldol) and chlorpromazine, have a side effect of suppressing movements.

Antidepressants, such as citalopram (Celexa), escitalopram (Lexapro), fluoxetine (Prozac, Sarafem) and sertraline (Zoloft).

Speech Therapy Occupational Therapy Provides a safe environment (jerking movement) Provides a calm and stress-free environment Provides ample time for communication Breaking down tasks into manageable steps Regular toileting intervals and skin care Monitoring nutritional intake; may need gastrostomy feedings Assist with ADLs Explain need for guardianship and advanced directives Use of hand rail while in home Use of assistive devices (ie. walker) Use of calendar and schedule to help keep routine Appropriate coping mechanisms Family Planning to prevent passing disease to offspring Alternatives (adoption) Sperm or egg donation Use of invitro to select only gene negative embryos for implantation