NGN NCLEX 2026 | NEUROLOGICAL / GENETIC | VISUAL REFERENCE NO. 047

Huntington's Disease

An autosomal dominant, progressive neurodegenerative disorder marked by chorea, cognitive decline, and psychiatric symptoms — with high suicide risk and no cure.

SOURCE TRANSPARENCY: This topic is not present in Diane's uploaded personal notes. Content has been compiled from standard NGN NCLEX 2026 references — Saunders Comprehensive Review (8th ed.), ATI Medical-Surgical Review, Lippincott Manual of Nursing Practice, and current HDSA clinical guidelines.

01 Definition & Overview

FOUNDATION

- **Inherited, autosomal dominant** neurodegenerative disease — each child of an affected parent has a **50% chance** of inheriting the mutated gene.
- Caused by an abnormal **CAG trinucleotide repeat expansion** on the *HTT gene* (chromosome 4) → produces a toxic mutant huntingtin protein.
- Progressive destruction of the **basal ganglia** (especially the caudate nucleus and putamen) and cortex → motor, cognitive, and psychiatric decline.
- Typical onset: ages **30–50 years**; juvenile-onset is rare. No cure — care is supportive and palliative.
- Death usually occurs **15–20 years** after symptom onset, most often from **aspiration pneumonia, injury, or suicide**.

Why it matters on NCLEX: HD pulls together genetics, neuro assessment, psychiatric risk (suicide), nutrition, aspiration precautions, and end-of-life planning — a high-yield clinical judgment topic.

02 Pathophysiology — Step by Step

MECHANISM

STEP 1 — GENETIC DEFECT

Expanded CAG repeats on HTT gene

Normal: < 27 CAG repeats. HD: ≥ **40 repeats** → fully penetrant disease. The more repeats, the earlier the onset.

STEP 2 — TOXIC PROTEIN

Mutant huntingtin protein produced

The defective gene codes a misfolded protein that **aggregates inside neurons** and disrupts cell function.

STEP 3 — NEURONAL DEATH

Selective loss in the basal ganglia

Medium spiny neurons of the **caudate nucleus and putamen** degenerate first → loss of GABA (inhibitory neurotransmitter) and acetylcholine.

STEP 4 — DOPAMINE EXCESS

Unopposed dopaminergic activity

Loss of GABA inhibition → relative **dopamine excess** in the striatum → the involuntary, dance-like **chorea** movements.

STEP 5 — CORTICAL SPREAD

Frontal cortex degeneration

Progression to the cortex causes **dementia, impulsivity, depression, and psychosis**. Motor + cognitive + psychiatric triad emerges.

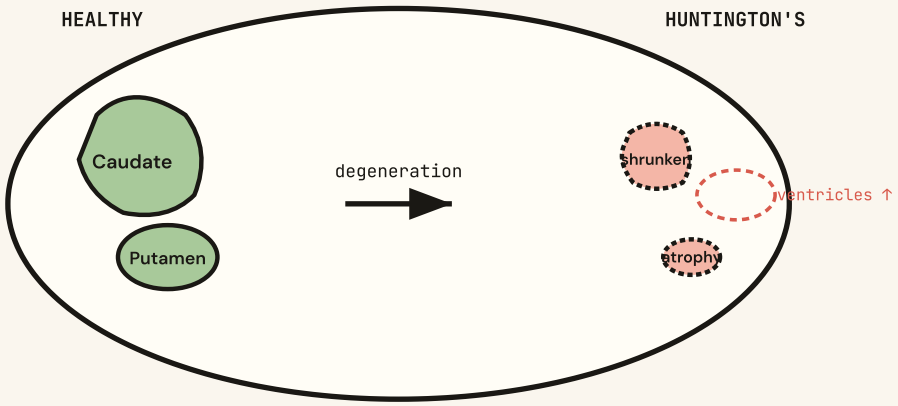

FIG. 1 — CAUDATE & PUTAMEN ATROPHY WITH COMPENSATORY VENTRICULAR ENLARGEMENT

03 Signs & Symptoms

ASSESSMENT

EARLY STAGE

Subtle & Easily Missed

- Personality changes — irritability, moodiness, apathy
- Mild depression or anxiety
- Clumsiness, dropping objects, restlessness
- Subtle fidgeting movements (often dismissed)
- Difficulty with multitasking and planning
- Mild memory lapses; insight is **preserved**

LATE STAGE

Severe & Life-Threatening

- **Severe chorea** — jerky, dance-like involuntary movements
- **Dysphagia** → aspiration pneumonia (#1 cause of death)
- Dysarthria — slurred, difficult speech
- Progressive **dementia**, disorientation
- Severe weight loss despite normal/↑ caloric intake
- Rigidity, immobility, total dependence
- Psychiatric: psychosis, paranoia, **suicidal ideation**
- Incontinence, contractures, pressure injuries

Clinical pearl — the HD triad: *Motor + Cognitive + Psychiatric.* Suicide risk peaks **early-to-mid stage**, when patients still have insight into their decline. Always screen.

04 Priority Nursing Interventions

ABC + SAFETY

A — AIRWAY

- **Assess swallow** before any PO intake
- Thickened liquids; soft, moist foods
- Upright (90°) for meals; remain upright 30 min after
- Suction at bedside
- Small bites, slow pace, chin-tuck

B — BREATHING

- Monitor lung sounds q shift — crackles = pneumonia risk
- Encourage coughing & deep breathing
- Track O_2 sat, RR, temperature
- Vaccinations: pneumococcal, influenza

C — CIRCULATION & SAFETY

- **Fall precautions** — bed low, padded environment
- Helmet/pads if severe chorea
- Soft restraints only if absolutely needed
- Skin assessment q shift (pressure injury)

NUTRITION (HIGH PRIORITY)

- **4,000–6,000 kcal/day** — chorea burns enormous calories
- High-calorie, high-protein, soft-texture diet
- Frequent small meals + nutritional supplements
- Weigh daily; monitor I&O
- Anticipate PEG tube as disease progresses

PSYCHIATRIC & SUICIDE RISK

Screen for suicidal ideation at **every visit**

- Assess depression, hopelessness, mood changes
- Refer to mental health; involve family
- Remove access to means; ensure supervision

Don't forget: Genetic counseling for the patient AND family, advance directives discussion early (while cognition intact), referral to HDSA and palliative care.

05 Pharmacology

SYMPTOMATIC ONLY

There is no cure or disease-modifying drug. All medications treat **symptoms**.

DRUG / CLASS	MECHANISM & USE	KEY SIDE EFFECTS	NURSING CONSIDERATIONS
Tetrabenazine (Xenazine) *Deutetrabenazine (Austedo)* VMAT2 inhibitor	Depletes dopamine in nerve terminals → **reduces chorea**. FDA-approved specifically for HD chorea.	🚨 **Depression & suicidality**, sedation, parkinsonism, akathisia, QT prolongation	Black box: monitor for **suicidal thoughts**; baseline ECG; avoid in active suicidality or untreated depression
Haloperidol **Risperidone, Olanzapine** Antipsychotics	Dopamine blockade → reduces chorea, psychosis, agitation	EPS, tardive dyskinesia, **neuroleptic malignant syndrome**, sedation, weight changes	Monitor for EPS, fever + rigidity (NMS), QT, falls
Fluoxetine, Sertraline SSRIs	Treat depression, anxiety, obsessive features	GI upset, sexual dysfunction, hyponatremia, ↑ suicide risk early	Reassess suicide risk; takes 2–4 wk for full effect
Clonazepam, Lorazepam Benzodiazepines	Anxiety, agitation, dystonia, sleep	Sedation, falls, dependence, respiratory depression	Fall precautions; lowest effective dose; avoid abrupt stop
Amantadine	NMDA antagonist — adjunct for chorea	Livedo reticularis, confusion, anticholinergic effects	Renal dosing; monitor mental status in elderly

06 NCLEX Test Traps ⚠

WRONG VS. RIGHT

✕ WRONG THINKING	✓ RIGHT THINKING
"HD is autosomal recessive — only happens if both parents carry it."	HD is **autosomal dominant**. One affected parent = **50% risk** for each child. Only one copy of the defective gene is needed.
"Patient has lost 20 lbs — they need a calorie-restricted diet because they can't exercise."	Chorea burns **thousands of calories daily**. HD patients need **4,000–6,000 kcal/day** of high-calorie, high-protein, soft food.
"Suicide risk is highest in late-stage HD when patients are bedbound."	Suicide risk peaks in **early to mid-stage** HD when patients retain insight into their decline. Screen at **every** visit.
"Huntington's and Parkinson's are basically the same — both cause tremors."	**Parkinson's** = rigidity, bradykinesia, resting tremor, dopamine *deficit*. **HD** = chorea (hyperkinetic), dopamine *excess* relative to GABA loss.
"Tetrabenazine cures HD by stopping the gene."	Tetrabenazine only **controls chorea**. It carries a **black box warning for depression and suicidality** and does not slow disease progression.
"Wait until late-stage to discuss advance directives — patient is still functional."	Discuss **advance directives early**, while cognition is intact. Decision-making capacity diminishes as the disease progresses.

07 Patient & Family Education

TEACHING

Genetic & Lifespan

- Each biological child has a **50% chance** of inheriting HD
- Predictive testing is available — recommend genetic counseling first
- Family planning options: PGD, adoption, donor gametes
- Connect family with the **Huntington's Disease Society of America (HDSA)**

Safety at Home

- Remove rugs, sharp corners, clutter; install grab bars
- Padded furniture and helmets if chorea is severe
- Lock up medications, firearms, vehicles
- Supervised meals; emergency response system

Nutrition & Swallowing

- High-calorie, high-protein, soft-texture foods
- Thickened liquids per speech therapy
- Upright posture for meals + 30 min after
- Discuss **PEG tube** proactively, not as a crisis

Mental Health & Support

- Report any mood changes, hopelessness, suicidal thoughts *immediately*
- Caregiver burnout is real — schedule respite care
- Join HD support groups (in-person or online)
- Plan early: advance directives, healthcare proxy, POLST

08 Memory Boosters & Mnemonics

PATTERN RECOGNITION

HUNTING

- **H** **H**ereditary — autosomal dominant (50%)
- **U** **U**ncontrolled movements (chorea)
- **N** **N**eurodegeneration of basal ganglia
- **T** **T**riad: motor + cognitive + psychiatric
- **I** **I**nevitable progression — no cure
- **N** **N**utrition needs are HIGH (chorea = calories)
- **G** **G**enetic counseling for the whole family

Pattern: "Dance of Death"

Chorea comes from the Greek for *"dance"*. Picture jerky, dance-like movements that the patient cannot stop — that's the hallmark motor sign of HD.

Quick Compare: HD vs. Parkinson's

HD = too much movement (hyperkinetic), dopamine excess. **Parkinson's** = too little movement (hypokinetic), dopamine deficit. *Opposite ends of the same axis.*

Lab Threshold

≥ 40 CAG repeats = full penetrance HD. Normal is < 27. The longer the repeat, the earlier the symptom onset.

Cause of Death — Top 3

① **Aspiration pneumonia** ② **Suicide** ③ **Injury from falls**. All three are nursing-preventable with priority interventions.

09 NCJMM Clinical Judgment Walkthrough

6-STEP MODEL

SCENARIO

A **42-year-old male** with confirmed Huntington's disease (CAG repeat = 44) is admitted from home for evaluation of a **20-lb weight loss over 3 months**, a recent fall, and increasing depression. His father died of HD at age 58. He is taking tetrabenazine and sertraline.

Vitals: T 99.4°F · HR 102 · RR 22 · BP 108/68 · SpO_2 94% on RA · Wt 138 lb (down from 158 lb).

Assessment: Continuous chorea of upper extremities, dysarthric speech, coughs after sips of water, denies SI but wife reports he said *"I don't want to end up like Dad."* Skin intact, gait unsteady.

STEP 1 — RECOGNIZE CUES 01

Identify relevant findings: significant **weight loss**, **coughing on liquids** (dysphagia), **SpO_2 94%**, low-grade fever and tachypnea (early aspiration?), unsteady gait + recent **fall**, statement suggesting **passive suicidal ideation**, ongoing chorea with high metabolic demand.

STEP 2 — ANALYZE CUES 02

Cluster the findings: ① **Aspiration risk** — dysphagia + low SpO_2 + tachypnea + low-grade fever. ② **Nutritional deficit** — 20-lb loss; chorea is burning thousands of kcal/day. ③ **Safety risk** — fall + chorea + unsteady gait. ④ **Psychiatric risk** — sertraline therapy + statement about not wanting to "end up like Dad" + tetrabenazine (black-box for suicidality).

STEP 3 — PRIORITIZE HYPOTHESES 03

Top priorities (by ABC + safety): ① **Aspiration / impaired airway** (Airway/Breathing — most immediately life-threatening). ② **Suicide risk** (life-threatening; both medications carry risk). ③ **Malnutrition**. ④ **Fall / injury risk**.

STEP 4 — GENERATE SOLUTIONS 04

Airway/Breathing: NPO until swallow study; suction at bedside; HOB ≥ 30°; speech therapy consult; pulse ox monitoring. **Suicide:** formal screening (C-SSRS); 1:1 supervision; remove means; psych consult; reassess tetrabenazine. **Nutrition:** dietitian; high-cal soft diet; daily weights; consider PEG. **Safety:** bed low, padding, assisted ambulation, PT consult.

STEP 5 — TAKE ACTION 05

First action: Place patient NPO and notify provider for swallow evaluation and ABG. **Next:** Complete formal suicide assessment and initiate continuous observation if indicated; ensure 2 g O_2 NC available; obtain chest X-ray for suspected aspiration; consult dietary, speech, PT, and psychiatry; document teaching with wife about HD triad & safety planning.

STEP 6 — EVALUATE OUTCOMES 06

Expected: No aspiration events; SpO_2 ≥ 95% on RA; safe swallow plan in place; weight stabilizes within 1 week; suicide risk assessed and trending lower; no falls. **Reassess if:** fever ≥ 100.4°F, new crackles, new SI, further weight loss, or chorea worsens — escalate to provider.