AAVBC

AMERICAN ACADEMY OF VALUE BASED CARE

Huntington's Disease

Quick Reference Guide

2026

Table of Contents

1 CLINICAL SNAPSHOT

Definition: Huntington's disease (**HD**) is an autosomal-dominant neurodegenerative disorder caused by CAG trinucleotide expansion in the huntingtin (**HTT**) gene.[1,2] The triad of **motor** (chorea, dystonia, bradykinesia), **cognitive** (executive dysfunction, dementia), and **psychiatric** (depression, irritability, apathy, psychosis) manifestations defines the **clinical syndrome**.[1,2] Neuropsychiatric and cognitive symptoms typically **precede** motor onset by **10–15 years**,[2] and late-onset HD (**LoHD**; symptom onset **≥60 years**) often presents with **gait instability** rather than classic chorea → frequently **misattributed** to Parkinson disease, vascular parkinsonism, or Alzheimer disease.[3]

ICD-10 Codes: Primary: **G10** (Huntington's disease). Cognitive manifestations: **F02.A0**–**F02.A4** (mild), **F02.B0–F02.B4** (moderate), **F02.C0–F02.C4** (severe) = **code G10 FIRST**;[4] **G31.84** (mild NCD) alone: do **NOT** add G10.[4] **Psychiatric manifestations** (etiologically linked to HD): **F06.31/ F06.32** (mood disorder due to physiological condition), **F06.4** (anxiety), **F06.0/F06.2** (psychotic features) = use physiological etiology codes, **NOT** primary psychiatric codes.[4] **Motor and somatic**: **R25.8** (chorea), **G24.8** (other dystonia), **R13.12** (oropharyngeal dysphagia), **J69.0** (aspiration pneumonia), **R26.81** (unsteadiness on feet), **R63.4** (abnormal weight loss); **Z82.0**/ **Z15.89** for family history and gene-positive pre-manifest status.[4]

Prevalence & Burden: US Medicare and Medicaid HD prevalence is **13.1 per 100,000** beneficiaries with incidence **6.1 per 100,000** person-years.[1] **LoHD** accounts for approximately **11–25%** of all HD and is growing; the proportion of new HD diagnoses with late onset rose from **21%** (1991–2000) to **33%** (2001–2010).[3] Notably, **31–53% of LoHD patients** have **no known family history** and this is the single most important driver of **missed diagnosis**.[3] **Aspiration pneumonia** is the leading cause of HD death (~19.5-22% of HD deaths);[5] Black patients are less than **half as likely** as White patients to be referred for genetic evaluation (OR 0.49, $p<0.001$),[6] and patients in the lowest-wealth quartile are significantly less likely to undergo genetic evaluation (OR 0.67).[7]

HCC V28/RAF Mapping

ICD-10 CODES[4,6]	HCC CATEGORY[7]	RAF WEIGHT (CNA)[8]	APPLICABILITY AND DOCUMENTATION[4]
G10 (Huntington Disease)	**HCC 200:** Friedreich and Other Hereditary Ataxias; Huntington Disease	0.279	Confirmed diagnosis by genetic testing (**CAG ≥36**) **OR** clinical motor DCL 4 with family history; document CAG repeat length in problem list **annually**; G10 must appear (primary or secondary) at **every encounter** for HCC 200 mapping
F02.C0–F02.C4 (major NCD due to HD, severe)	**HCC 125:** Dementia/Severe	0.341	Major NCD due to HD, **severe**; advanced-stage HD with documented severe NCD; code G10 **FIRST** and **explicitly state** NCD severity

ICD-10 CODES[4,6]	HCC CATEGORY[7]	RAF WEIGHT (CNA)[8]	APPLICABILITY AND DOCUMENTATION[4]
F02.B0–F02.B4 (major NCD due to HD, moderate)	**HCC 126:** Dementia/ Moderate	0.341	Major NCD due to HD, **moderate severity**; document behavioral/ psychotic/mood/anxiety disturbance subtype when present; severity must be **explicitly stated**
F02.A0–F02.A4 (major NCD due to HD, mild)	**HCC 127:** Dementia/ Mild or Unspecified	0.341	Major NCD due to HD, **mild severity**; code G10 **FIRST**; document behavioral/ psychotic/mood/anxiety disturbance subtype when present; severity must be **explicitly stated**
F02.80, F02.811, F02.818, F02.82–F02.84 (major NCD due to HD, unspecified severity)	**HCC 127:** Dementia/ Mild or Unspecified	0.341	**Unspecified severity NCD**; upgrade to severity-specific codes (**F02.A–F02.C**) when documentation supports; wrongly assigned unspecified code (where severity is documented) = **coding compliance gap**
F06.4, F06.31 (anxiety and mood disorders due to known physiological condition)	None	—	Anxiety or mood disturbance **directly attributable to HD**; supports clinical complexity documentation even without independent HCC assignment
G31.84 (mild neurocognitive disorder due to known physiological condition)	None	—	Mild NCD due to HD; do **NOT** add G10 per DSM-5-TR, **G31.84 stands alone**;[4] lower coding weight than F02.x; chart review required to confirm NCD **has not progressed** to major **before** assigning
G24.8, R25.8, R13.12, R26.81, R63.4 (motor/ somatic manifestations)	No direct HCC (symptomatic codes)	—	Code **each** symptomatic manifestation linked to HD (dystonia, chorea, dysphagia, unsteadiness, weight loss); supports clinical complexity documentation even without independent HCC assignment

ABBREVIATIONS: ICD-10 = International Classification of Diseases, 10th Edition; HCC = hierarchical condition category; V28 = CMS-HCC model version 28; RAF = risk adjustment factor; CNA = community non-dual aged; CAG = cytosine-adenine-guanine (trinucleotide repeat); DCL = diagnostic confidence level; NCD = neurocognitive disorder; HD = Huntington disease; DSM-5-TR = Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition, Text Revision; MEAT = monitor/evaluate/assess/treat; CMS = Centers for Medicare & Medicaid Services

RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting

Risk-Adjusted Care Resources per Patient/Year[6,7]

Risk-adjusted care resource allocation — MA base rate ($10,402.34) × RAF coefficient

HD	HD + NCD
$2.9K	**(severe/moderate/mild/ unspecified)**
HCC 200 · RAF 0.279	$6.5K+

RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting

AAVBC PERSPECTIVE

***From the AAVBC's perspective**, value-based care in Huntington's disease demands a fundamental shift in clinical attention: **proactively screen for and manage non-motor symptoms** (psychiatric and cognitive changes) **before** chorea becomes prominent. HD neuropsychiatric symptoms are neurodegenerative, not reactive, and carry the **same clinical urgency** as motor manifestations:[3,9] HD confers approximately a **5-fold increase in suicide risk** compared to the general population.[10,11] When a patient presents with treatment-resistant depression, irritability, apathy, or executive dysfunction, **even without motor signs**, a thorough three-generation family history focused on neurological and psychiatric disease should be obtained. **Any family member** with an unexplained movement disorder, early dementia, psychiatric illness, or early death should **raise clinical suspicion significantly**; up to **31%** of late-onset HD cases lack a known family history because the affected relative was **never diagnosed or died before presentation**.[12] Clinicians must also recognize that medications commonly prescribed in elderly patients can both **mimic early HD** (**tardive dyskinesia** from antipsychotics resembling chorea; drug-induced parkinsonism resembling late-stage rigidity) and **mask HD** (anticholinergics **obscuring cognitive decline**; benzodiazepines blunting executive dysfunction). A patient on risperidone who develops **rigidity and gait instability** may have symptoms attributed entirely to drug effects when they are the **first signs of HD** superimposed on those effects. **AAVBC emphasizes** that primary care is uniquely positioned to close the diagnostic gap in late-onset HD by integrating psychiatric screening, medication reconciliation, and family history assessment into routine encounters for older adults with unexplained neuropsychiatric decline, ensuring that **treatment decisions incorporate**: functional status, caregiver dynamics, polypharmacy burden, and patient goals alongside disease biology.*

2 RECOGNITION AND DIAGNOSIS

In older adults (onset >60 years), HD overwhelmingly presents with **motor abnormalities first** (~69% of cases), but cognitive onset may be **underrecognized** because it is attributed to age-related change, making it essential to assess across the **5Ms** (Mind, Mobility, Medications, Multi-complexity, Matters Most) when chorea, gait unsteadiness, executive decline, or new-onset irritability/depression **co-occur** with a **positive family history**.[1,3] Confirmation requires **HTT CAG-repeat genetic testing** (fundamental diagnostic test per ACMG 2014/2021)[13,14] integrated with structured cognitive (**MoCA**) and psychiatric (**PHQ-9**/**C-SSRS**) assessment to stage severity, drive ICD-10 code selection, and establish the longitudinal monitoring baseline.

Diagnostic Workups Covered under Medicare Part B (symptom- or family-prompted)

TEST	FREQUENCY	CPT CODE	CLINICAL INDICATION
HTT CAG-repeat genetic testing	**Once per lifetime***	81271	**≥40** repeats = **full penetrance**, **>99%** sensitivity for symptomatic disease; **36–39** repeats = **reduced penetrance** → relevant to Medicare population; pre- and post-test genetic counseling required per ACMG 2014/2021[13,14]
UHDRS motor exam (DCL 4)	**At suspicion**; annually thereafter	99203 – 99215	Clinical motor **DCL 4** + family history confirms HD **without genetic testing** in some settings; integrated into E/M neurology visit[15]
MoCA/cognitive assessment	**At baseline** + annually	96125/ 99483	Document **cognitive trajectory**; supports F02.x severity coding and **HEDIS COA** cognitive measure;[4] covered under Part B for cognitive symptoms
PHQ-9/C-SSRS suicidality screen	**Every visit**†	96127/ G0444	HD-related depression carries **2–7x** general-population suicide risk;[5,16] screen at **every PCP visit**; USPSTF **Grade B** recommendation for depression screening[17]
FEES (dysphagia assessment)	**At symptom onset**; per progression‡	92612/ 92613	**Silent aspiration** occurs in **7.7–27.8%** of HD patients across stages;[18] SLP referral **proactively**, not reactively; instrumental assessment recommended over clinical bedside assessment alone[19]

TEST	FREQUENCY	CPT CODE	CLINICAL INDICATION
Advance care planning (AWV)	**Annually** + when capacity changes§	99497, 99498	Initiate while **capacity intact**; HD-specific complexity from gene-positive proxies; no copay when billed during AWV

ABBREVIATIONS: HTT = huntingtin gene; CAG = cytosine-adenine-guanine (trinucleotide repeat); CPT = Current Procedural Terminology; ACMG = American College of Medical Genetics; UHDRS = Unified Huntington Disease Rating Scale; DCL = diagnostic confidence level; HD = Huntington disease; E/M = evaluation and management; MoCA = Montreal Cognitive Assessment; HEDIS = Healthcare Effectiveness Data and Information Set; COA = cognitive assessment; PHQ-9 = Patient Health Questionnaire-9; C-SSRS = Columbia Suicide Severity Rating Scale; PCP = primary care provider; USPSTF = United States Preventive Services Task Force; FEES = fiberoptic endoscopic evaluation of swallowing; SLP = speech-language pathology; AWV = annual wellness visit; ACP = advance care planning; LCD = local coverage determination; MA = Medicare Advantage; PA = prior authorization
***Genetic testing: Part B covers when medically necessary (signs/family history); no population screening; LCD-variable; MA plans may require PA**
† Depression screening: Part B–covered per USPSTF Grade B; every-visit frequency reflects HD-specific suicide risk
‡ FEES: Part B covers for documented dysphagia only
§ ACP: Part B–covered; copay waived during AWV

Subtle Early Signs in Older Adults (Late-Onset HD)

SIGN/SYMPTOM[2,3,16]	CLINICAL SIGNIFICANCE
Treatment-resistant depression with executive dysfunction	HD neuropsychiatric symptoms **precede** motor onset by **10–15 years**;[2] **31–53%** of LoHD patients lack known family history;[2,3] suspect HD when **standard antidepressants fail** and cognitive symptoms emerge
Subtle motor abnormalities: eye-tracking deficits, mild involuntary movements, soft chorea	LoHD often presents with gait instability and balance problems **rather than classic chorea**;[3] examine eye saccades, slowed initiation, and finger tapping in patients with **unexplained psychiatric symptoms**
Apathy distinct from depression	**Apathy** (loss of motivation without sadness) responds poorly to SSRIs → treatment differs from depression;[16] environmental structuring and activity scheduling are **first-line**
Unexplained gait instability or recurrent falls	Gait instability is the **predominant presenting feature** in LoHD;[3] **misattributed** to age, Parkinson disease, or vascular parkinsonism without HD evaluation
New-onset chorea in patient ≥60 on dopamine receptor blocking agents	TD and HD chorea can **coexist or mimic each other**;[4] TD prevalence 13% (SGAs) to 32% (FGAs), with older age the strongest risk factor;[20] HD is distinguished from TD by insidious onset, progressive cognitive decline, motor impersistence, and impaired saccades; **DaT-SPECT** differentiates drug-induced from neurodegenerative parkinsonism when parkinsonian features overlap[21]

SIGN/SYMPTOM[2,3,16]	CLINICAL SIGNIFICANCE
Family history of neurodegenerative disease	Patient **may not know** HD-specific diagnosis but may report **family history of:** 'dementia,' 'movement disorder,' early death, or psychiatric illness;[2] LoHD often emerges in families **without prior recognized HD**

ABBREVIATIONS: HD = Huntington disease; LoHD = late-onset Huntington disease; SSRIs = selective serotonin reuptake inhibitors; TD = tardive dyskinesia; SGAs = second-generation antipsychotics; FGAs = first-generation antipsychotics; DaT-SPECT = dopamine-transporter single-photon emission computed tomography

Geriatric Risk Factors

FACTOR[2,3]	RISK SIGNAL	NOTES
Age ≥60 with prodromal psychiatric or motor symptoms	LoHD accounts for ~11% of HD in the Enroll-HD cohort[12]	**Up to 31% of LoHD lacks known family history;**[22] **consider genetic testing in elderly with treatment-resistant depression and subtle motor signs**
Family history of any neurological/psychiatric disease	Fewer LoHD patients have **positive family history** vs common-onset HD (p0.001)[22]	Family terms like 'dementia,' 'movement disorder,' or 'mental illness' **may signal HD**; low threshold for **genetic counseling referral**[23]
Antipsychotic or dopamine-blocking drug exposure	TD prevalence **~30%** in patients on first-generation antipsychotics[24]	**TD mimics HD chorea**; DaT-SPECT can distinguish **drug-induced** from **neurodegenerative parkinsonism**
Cumulative VMAT2 inhibitor exposure in elderly	**Tetrabenazine AEs** per AAN guideline: sedation **24%**, depressed mood **23%**; FDA label: parkinsonism **9%***[25]	Reassess need as **chorea wanes in late HD**; monitor for falls, depression, and sedation*[25]
Black race/lower-wealth quartile	In a US cohort, Black patients **half as likely** to be referred for genetic evaluation (OR 0.49); **lowest-wealth quartile** also less likely (OR 0.67)[26]	Proactively offer genetic counseling; address access barriers
Rural setting	Rural neurology access **~80% lower** than metropolitan in the US[27]	Use **telehealth genetic counseling** and EHR-based screening prompts to close the gap
Polypharmacy with anticholinergic burden	Cumulative strong anticholinergic use linked to **increased dementia risk** (HR 1.54) in a US cohort[28]	Review anticholinergic load at every visit per **AGS Beers Criteria**;[29] switch agents where possible

FACTOR[2,3]	RISK SIGNAL	NOTES
Caregiver who may also be HD gene-positive	HD is autosomal dominant = 50% offspring risk	Flag potential conflict of interest when **proxy is gene-positive**; **refer family** for genetic counseling

ABBREVIATIONS: LoHD = late-onset Huntington disease; HD = Huntington disease; TD = tardive dyskinesia; DaT-SPECT = dopamine-transporter single-photon emission computed tomography; VMAT2 = vesicular monoamine transporter 2; AEs = adverse events; AAN = American Academy of Neurology; FDA = Food and Drug Administration; OR = odds ratio; EHR = electronic health record; HR = hazard ratio; AGS = American Geriatrics Society
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

HD Diagnosis, Staging and Classification

Confirming Huntington disease requires **HTT CAG-repeat molecular genetic testing** (CPT 81271), which defines allele status and penetrance.[1] Once confirmed, two complementary clinical staging systems guide management: the **UHDRS Diagnostic Confidence Level (DCL)** classifies motor certainty, and the **Shoulson-Fahn Total Functional Capacity (TFC) scale** grades functional severity into five stages.[30,31]

The **HD Integrated Staging System (HD-ISS)** provides a newer biological research framework enabling classification **before motor diagnosis**.[2] Proposed 2025 diagnostic criteria also now recognize nonmotor-onset HD, allowing clinical diagnosis in a confirmed genetic carrier based on persistent cognitive, behavioral, or psychiatric syndromes **without unequivocal motor signs**.[15] Late-onset HD (onset ≥60 years) comprises ~11% of cases, presents predominantly with motor symptoms (~69%), and up to 31% lack a known family history, making the diagnosis easily missed or attributed to age-related neurodegeneration.[12,32] Pre- and post-test genetic counseling is **required** per ACMG guidelines regardless of whether testing is **presymptomatic** or **clinically motivated**.[13,14,23,33]

HTT CAG-Repeat Allele Classification (ACMG 2014/2021)

CAG REPEAT RANGE[13,14]	CLASSIFICATION[13,1]	CLINICAL IMPLICATION[13,14,33]
≤26	Normal	**No HD risk**; no risk to offspring
27–35	Intermediate (mutable normal)	**No HD phenotype**; meiotically unstable → **offspring may inherit** expanded allele in disease-causing range
36–39	Reduced penetrance	**May or may not develop HD**; elderly asymptomatic carriers in this range are **common**; ~one-third carry noncanonical sequence variants (CAG-CCG LOI) that **accelerate onset by ~10 years** = **undetectable** by standard PCR
≥40	Full penetrance	**HD expected** given normal lifespan; **>99%** sensitivity for symptomatic disease; CAG length inversely correlated with age of onset (~60% of onset variance)

ABBREVIATIONS: CAG = cytosine-adenine-guanine (trinucleotide repeat); HD = Huntington disease; CCG = cytosine-cytosine-guanine (trinucleotide repeat); LOI = loss of interruption; PCR = polymerase chain reaction

UHDRS Diagnostic Confidence Level (DCL)

DCL[15,30,34]	MOTOR CERTAINTY[15,30,34]	CLINICAL IMPLICATION[15,30,34]
0	Normal	**No** motor abnormalities
1	50% confidence	Nonspecific motor impairment; **does not suggest HD**
2	50-89% confidence	Motor impairment that **may be a sign of HD**; **prodromal stage**
3	90-98% confidence	Motor impairment **likely a sign of HD**; manifest HD if **cognitive changes significant** and evidence of progression
4	≥99% confidence	**Unequivocal motor signs of HD**; historically defines "**manifest**" **HD** and clinical motor diagnosis

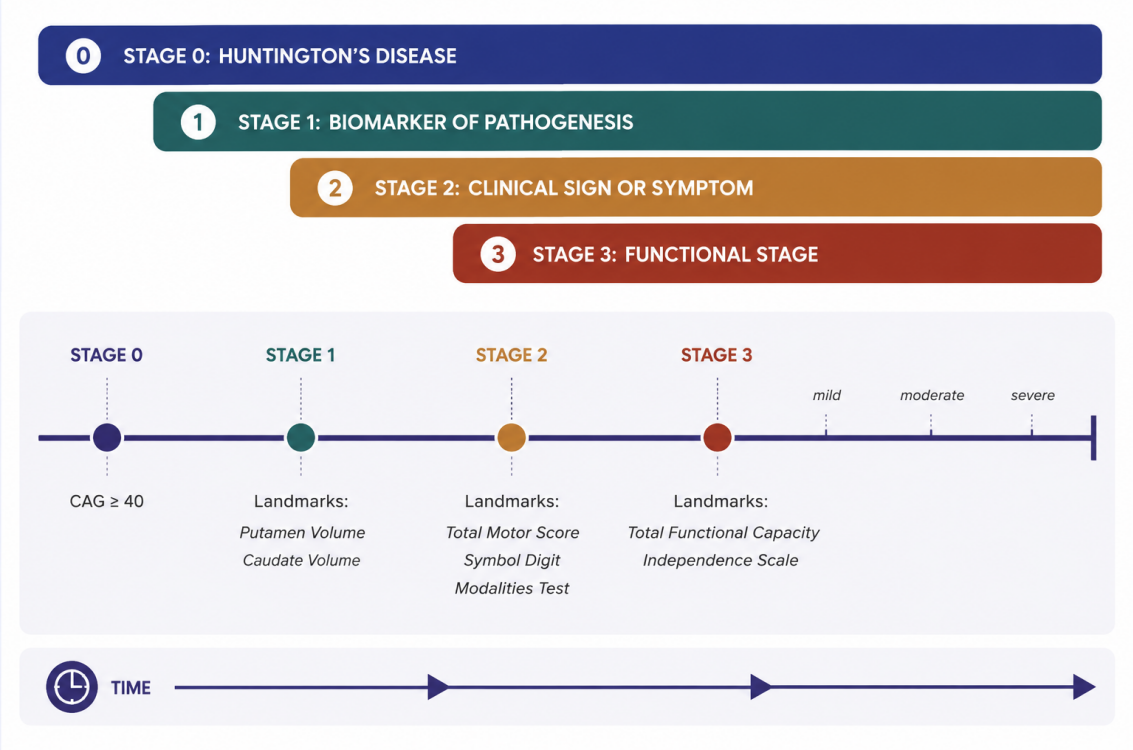
ABBREVIATIONS: HD = Huntington's disease

Shoulson-Fahn Functional Staging (TFC Scale, 0–13)

STAGE[31]	TFC SCORE[30,31]	STATUS[30,31]	CLINICAL IMPLICATION[30,31,35]
I	**11–13**	Early; fully functional	**Employed**; manages finances and ADLs **independently**; may have **subtle** motor/cognitive signs
II	**7–10**	Early-intermediate	**Reduced work capacity**; manages most ADLs; **mild chorea**; cognitive slowing detectable on testing
III	**4–6**	Intermediate	**Cannot work or manage finances**; requires assistance with some ADLs; moderate chorea; dysphagia risk increases
IV	**1–3**	Late-intermediate	Requires **substantial assistance**; cannot perform domestic chores; significant **motor and cognitive impairment**
V	**0**	Advanced	**Total care required**; severe motor disability; profound cognitive decline; **high aspiration risk**

ABBREVIATIONS: TFC = Total Functional Capacity; ADLs = activities of daily living

HD Integrated Staging System (HD-ISS) - Research Framework



ABBREVIATIONS: CAG = cytosine-adenine-guanine trinucleotide repeat expansion in the huntingtin (HTT) gene

Clues to Dig Deeper

CLINICAL CLUE	WHY IT MATTERS	NEXT DIAGNOSTIC STEP
Treatment-resistant depression + new executive dysfunction in patient ≥60	HD neuropsychiatric symptoms precede motor onset **by 15–20 years**; LoHD **lacks family history** in 31–53%[36]	**Neurology referral**; HTT genetic testing (**CPT 81271**) if any motor sign **or** family history of any neurological/ psychiatric disease[15]
Subtle eye-tracking deficits, slowed saccades, soft chorea in adult ≥60	LoHD presents predominantly with **gait instability** → chorea may be soft or absent[22]	**UHDRS motor exam**;[37] HTT genetic testing if **any** motor findings
Patient on antipsychotic with new movement disorder[9]	TD on first-generation antipsychotics **~32%**; can mimic OR mask HD chorea[13,14]	**DaT-SPECT** to distinguish DIP from neurodegenerative;[38] neurology referral; consider HTT testing
Apathy not responding to SSRI trials[11]	**Apathy ≠ depression** in HD; **SSRIs ineffective**[9]	**Environmental structuring**; reconsider HD evaluation if other features present

CLINICAL CLUE	WHY IT MATTERS	NEXT DIAGNOSTIC STEP
Family history of dementia, movement disorder, or early death without specific HD diagnosis	Many LoHD families **lack prior HD identification**;[36] family history is the **strongest screening trigger**	Genetic counseling referral; **pre-test counseling**[13,14]
Suicidal ideation in patient with motor or cognitive symptoms[1]	HD carries **~5x** general-population suicide risk[10]	C-SSRS **at every visit**; urgent psychiatry referral; **AVOID** tetrabenazine and VMAT2 inhibitors with untreated suicidality*

ABBREVIATIONS: HD = Huntington disease; LoHD = late-onset Huntington disease; HTT = huntingtin (gene); CPT = Current Procedural Terminology; UHDRS = Unified Huntington Disease Rating Scale; TD = tardive dyskinesia; DaT-SPECT = dopamine-transporter single-photon emission computed tomography; DIP = drug-induced parkinsonism; SSRI = selective serotonin reuptake inhibitor; C-SSRS = Columbia Suicide Severity Rating Scale; VMAT2 = vesicular monoamine transporter 2
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Common Oversights

OVERSIGHT/SHORTCUT	WHY IT MATTERS — WHAT TO DO INSTEAD
Withholding HD evaluation in patient with no known family history	31–53% of LoHD lacks known family history;[36] family history is **helpful** but its absence does **NOT** rule out HD
Continuing VMAT2 inhibitors as chorea naturally diminishes in late-stage rigid phase	VMAT2 inhibitors **worsen rigidity** in late HD;[39] **deprescribe** and pivot to fall prevention and comfort care
Initiating tetrabenazine or VMAT2 inhibitor without screening suicidality	VMAT2 inhibitors **contraindicated** in untreated/inadequately treated depression or suicidality; C-SSRS at **every visit**
Reactive (rather than proactive) dysphagia management	**Aspiration pneumonia** is the leading cause of HD death (19.5%);[5] FEES assessment, SLP, **modified diets**, and PEG discussions **BEFORE** crisis
Anticholinergic medication use in HD patient with cognitive symptoms	**Anticholinergic burden** masks early HD cognitive symptoms and **worsens existing impairment**;[40] review medication list at **every visit**

ABBREVIATIONS: HD = Huntington disease; LoHD = late-onset Huntington disease; VMAT2 = vesicular monoamine transporter 2; C-SSRS = Columbia Suicide Severity Rating Scale; FEES = fiberoptic endoscopic evaluation of swallowing; SLP = speech-language pathology; PEG = percutaneous endoscopic gastrostomy

Key Differentials in Elderly

PRESENTATION[9,12]	DIFFERENTIAL	KEY TESTS
Chorea or involuntary movement in patient ≥60	**LoHD vs.** TD; senile chorea; Wilson disease; thyrotoxicosis; SLE/antiphospholipid; cerebrovascular chorea[4]	Medication review; **HTT genetic testing**; TSH; ceruloplasmin if young-onset; brain MRI
Cognitive decline + movement disorder	**LoHD vs.** Alzheimer disease with parkinsonism; Lewy body dementia; vascular dementia with gait abnormality; corticobasal syndrome	MoCA/neuropsych; HTT testing; **DaT-SPECT** for parkinsonism distinction
Treatment-resistant depression with executive dysfunction	**LoHD vs.** bipolar depression; pseudodementia; frontotemporal dementia; B12 deficiency; hypothyroidism	**Family history**; HTT testing if motor signs;[12] B12, TSH; neuropsych testing
LoHD presenting with gait instability	**HD (LoHD) vs.** Parkinson disease; vascular parkinsonism; NPH; cerebellar ataxia	**UHDRS exam**;[36] brain MRI; CSF if NPH suspected; HTT testing
Patient on antipsychotic with new movement disorder	**TD vs.** drug-induced parkinsonism; **HD vs.** acute dystonia; akathisia; withdrawal dyskinesia[4]	**DaT-SPECT** to distinguish DIP from neurodegenerative parkinsonism (sensitivity/specificity up to **85%**);[41] neurology referral; **consider HTT testing**
Suicidal ideation in patient with motor or cognitive symptoms	**HD with suicidality vs.** major depressive disorder; bipolar disorder; substance use disorder; adjustment disorder[4]	C-SSRS screening at every visit;[10,42] **urgent psychiatry referral**; **AVOID** tetrabenazine/VMAT2 inhibitors with untreated depression

ABBREVIATIONS: LoHD = late-onset Huntington disease; TD = tardive dyskinesia; SLE = systemic lupus erythematosus; HTT = huntingtin (gene); TSH = thyroid-stimulating hormone; MRI = magnetic resonance imaging; MoCA = Montreal Cognitive Assessment; DaT-SPECT = dopamine-transporter single-photon emission computed tomography; SSRIs = selective serotonin reuptake inhibitors; NPH = normal-pressure hydrocephalus; UHDRS = Unified Huntington Disease Rating Scale; CSF = cerebrospinal fluid; DIP = drug-induced parkinsonism; C-SSRS = Columbia Suicide Severity Rating Scale; VMAT2 = vesicular monoamine transporter 2

Comorbidity Screening

CONDITION	PREVALENCE/ASSOCIATION	SCREENING APPROACH
Depression (F06.31/F06.32)	**Significantly elevated in HD**;[1] precedes motor onset by 10–15 years; suicide risk **~5x** general population[10]	**PHQ-9 at every visit**; C-SSRS if PHQ-9 Item 9 positive; SSRI first-line
Anxiety (F06.4)	Common in HD[1]	GAD-7 annually; SSRI per psychiatry
Apathy (no direct ICD-10)	Distinct from depression; **responds poorly to SSRIs**[16]	**Apathy Evaluation Scale**; environmental structuring first-line[16]
Dysphagia (R13.12/R13.10)	35% early-stage, 94% moderate, **100% advanced HD**;[43] silent aspiration **7.7–27.8%**	**FEES assessment**; SLP referral; **modified diet textures** (IDDSI)[43]
Aspiration pneumonia (J69.0)	**Leading cause of HD death** (~19.5%);[11] oropharyngeal dysphagia **OR 11.9 for pneumonia**	**Proactive dysphagia management**;[43] early PEG discussions while capacity intact
Weight loss/cachexia (R63.4, E46)	Progressive weight loss = **hallmark of HD**; TEE is 11–14% higher than controls due to involuntary movements and a hypermetabolic state;[44] weight loss **correlates with CAG repeat length** independently of motor severity[45]	Weight at **every visit**; nutrition therapy; caloric supplementation **by individual assessment**
Falls/unsteadiness (R26.81)	**High fall risk** from chorea, dystonia, gait instability; amplified by **VMAT2-induced parkinsonism**[39]	**Fall risk screening** (HOS); PT/OT referral; home safety review
Polypharmacy/ anticholinergic burden	Anticholinergics **mask cognitive symptoms**; benzodiazepines and neuroleptics carry Beers Criteria risk[29]	Medication reconciliation at **every visit**; deprescribe per Beers[29]

ABBREVIATIONS: HD = Huntington disease; PHQ-9 = Patient Health Questionnaire-9; C-SSRS = Columbia Suicide Severity Rating Scale; SSRI = selective serotonin reuptake inhibitor; GAD-7 = Generalized Anxiety Disorder-7; ICD-10 = International Classification of Diseases, 10th Edition; FEES = fiberoptic endoscopic evaluation of swallowing; SLP = speech-language pathology; IDDSI = International Dysphagia Diet Standardisation Initiative; OR = odds ratio; PEG = percutaneous endoscopic gastrostomy; TEE = total energy expenditure; CAG = cytosine-adenine-guanine; CPT = Current Procedural Terminology; VMAT2 = vesicular monoamine transporter 2; HOS = Health Outcomes Survey; PT = physical therapy; OT = occupational therapy

⚠ RED FLAG — EMERGENCY/ACTIVATE CRISIS RESPONSE

These emergency conditions must be **immediately addressed** for patients with Huntington disease:

- **Active suicidal ideation with plan**
  - → ED transfer; 1:1 observation; emergent psychiatric evaluation
  - × **Do NOT initiate tetrabenazine/VMAT2 inhibitors with active suicidality**
- **Aspiration event with respiratory distress**
  - → **ED transfer**; code **J69.0**; **NPO** pending swallowing reassessment by SLP[18]
- **Neuroleptic malignant syndrome on VMAT2 inhibitor or antipsychotic (rigidity, hyperthermia, autonomic instability)**
  - → **Discontinue causative agent immediately**; ED transfer
- **Acute psychosis with safety threat**
  - → Psychiatric emergency evaluation; coordinate with HD specialist; low-dose atypical antipsychotic[46]

⚠ RED FLAG — URGENT (24-72 HOURS)

These emergency conditions require **expedited evaluation** in patients with Huntington disease:

- **New suicidal ideation without plan**
  - → Same-day psychiatric evaluation; C-SSRS screening; medication review
  - × Avoid **VMAT2 inhibitor initiation** with untreated depression[46]
- **Acute dysphagia worsening or new aspiration risk**
  - → Urgent **FEES** and **SLP referral**; consider hold on oral diet pending assessment[43]
- **Rapid functional decline — TFC drop ≥2 in <6 months**
  - → Review for VMAT2-induced parkinsonism, undertreated depression, polypharmacy, dehydration
  - → Falls and drug-induced parkinsonism in older adults can precipitate permanent loss of independence[21,38]
- **New psychotic symptoms (hallucinations, paranoia, disorganized behavior)**
  - → Urgent psychiatric evaluation; low-dose atypical antipsychotic per HD specialist[9,16]

3 MEAT DOCUMENTATION ESSENTIALS

Incomplete documentation is preventable: coding **G10 alone** without separately coding its cognitive, psychiatric, and motor manifestations **fails to capture the clinical complexity** that drives care planning, quality measurement, and risk adjustment.[3,4,15,16] DSM-5-TR requires **G10 + F02.x** (**with severity tier**) for major neurocognitive disorder due to HD, and **F06.31**/**F06.4** for associated mood and anxiety disorders, **not** primary psychiatric codes.[4] **HCC 200** (G10) carries a RAF of **0.279**, but the dementia codes (**F02.A–C**) map to **HCCs 125–127** (RAF **0.341**), meaning undercoding the cognitive component leaves significant **clinical complexity undocumented**.[4,8] The MEAT framework below demonstrates how each element translates into a defensible, **clinically accurate encounter note**:

***Case vignette**: A 67-year-old patient with **treatment-resistant depression** × 4 years and recent **gait instability** presents for follow-up. Family history reveals a paternal aunt with "**early dementia**." UHDRS exam shows soft choreiform movements of fingers, slowed saccades, and mild executive dysfunction (**MoCA 22**). HTT genetic testing: CAG 43 repeats = **full penetrance**. Diagnosed with late-onset HD.*

MONITOR: TFC: "TFC 10/13 (Shoulson-Fahn Stage II = early/intermediate). **MoCA 22** (mild cognitive impairment). Weight stable 68 kg × 12 months; nutritional intake adequate; **no aspiration symptoms**. **C-SSRS**: positive ideation, no plan or intent → psychiatry consult placed. Medication review: **no antipsychotics or anticholinergics**; sertraline 100 mg/day continued."

EVALUATE: "UHDRS motor exam: soft choreiform movements, slowed saccades, DCL 4 with family history. HTT genetic testing: CAG 43 = full penetrance, >99% sensitivity for symptomatic disease. Brain MRI: mild caudate atrophy. FEES: **no aspiration**; safe oral diet, baseline established for serial monitoring (silent aspiration present in 7.7% of early-stage HD)."

ASSESS: "Active Huntington disease (late-onset, Stage II), Shoulson-Fahn TFC 10: **G10**. Associated: mild neurocognitive disorder due to HD, **G31.84 alone** (do NOT add G10 per DSM-5-TR). Depressive disorder due to HD = F06.31 (NOT primary F32.x). Chorea: **R25.8** documented as HD-attributed."

TREAT: "Sertraline 100 mg/day continued for depression; C-SSRS at **every visit** given elevated suicide risk. Chorea: not yet functionally impairing → **defer VMAT2 inhibitor** (deutetrabenazine or valbenazine → initiate when chorea **impairs function or safety**). Specialist team: HD-specialist neurology, genetic counseling for family implications, psychiatry, SLP for proactive dysphagia surveillance, PT/OT for **fall prevention**. Advance care planning: CPT 99497; initiated while capacity intact; surrogate decision-maker discussion with awareness that family proxies **may also be gene-positive**."

Clinical Documentation Elements

- **Code G10 first, then manifestations:** DSM-5-TR mandates **G10 FIRST** then **F02.x** for major NCD due to HD;[4] severity tier (mild **F02.A0–A4**, moderate **F02.B0–B4**, severe **F02.C0–C4**) supports **correct HCC** (127, 126, 125)[8]
- **Mild NCD = G31.84 alone:** For mild neurocognitive disorder due to known physiological condition, code **G31.84 ALONE** = do **NOT** add **G10** per DSM-5-TR[4]

- **Psychiatric manifestations use F06.x:** F06.31/F06.32 for **mood**, F06.4 for **anxiety**, F06.0/F06.2 for **psychotic features**;[4] primary **F32.x**/**F41.x miss** the HD etiologic link
- **Document motor and somatic manifestations: R25.8** (chorea), **G24.8** (dystonia), **R13.12** (oropharyngeal dysphagia), **J69.0** (aspiration), **R26.81** (unsteadiness), **R63.4** (weight loss)[4]
- **Document TFC and CAG length:** Total Functional Capacity (**TFC 0–13**) anchors **disease staging**; CAG repeat length supports the genetic diagnosis

Reframing Common Documentation Shortcuts

INSTEAD OF…	DOCUMENT…[1,4]
'Huntington's disease'	'Huntington's disease (**G10**), Stage II per Shoulson-Fahn **TFC 10**; **CAG 43 repeats** (full penetrance) confirmed [date]'
'Dementia in HD' (without severity)	'Major neurocognitive disorder due to HD, moderate severity with behavioral disturbance (**F02.B11**);' coded **after G10** per DSM-5-TR
'Depression' in known HD patient	'Mood disorder due to HD with major depressive-like episode (**F06.32**);' NCD **etiologically linked** to HD (**G10**), **NOT** primary F32.x
'Chorea, on Austedo'	'Chorea due to HD (**R25.8 + G10**) Stage II, **functionally impairing**; deutetrabenazine 36 mg/day with **monthly C-SSRS suicidality screen** and depression monitoring per FDA label'
'Dysphagia' (HD patient)	'Oropharyngeal dysphagia due to HD (**R13.12 + G10**), FEES [date] confirmed pharyngeal-phase deficit; **modified texture diet** IDDSI level 4 with thickened liquids; SLP managing'
'History of HD' (in symptomatic patient)	'Active HD (**G10**) per UHDRS motor exam and CAG 43;' **never** use Z85.07-equivalent/Z82.0 history codes for active HD

ABBREVIATIONS: TFC = Total Functional Capacity; CAG = cytosine-adenine-guanine; HD = Huntington disease; NCD = neurocognitive disorder; DSM-5-TR = Diagnostic and Statistical Manual of Mental Disorders, 5th Edition, Text Revision; C-SSRS = Columbia Suicide Severity Rating Scale; FDA = US Food and Drug Administration; FEES = fiberoptic endoscopic evaluation of swallowing; IDDSI = International Dysphagia Diet Standardisation Initiative; SLP = speech-language pathology; UHDRS = Unified Huntington Disease Rating Scale

DOCUMENTATION IS COMPREHENSIVE CODING

*Every HD presentation is **patient-specific:** a distinct, multifaceted combination of motor, psychiatric, and cognitive features that evolves across the disease trajectory. **At every active HD visit**, this individuality must be reflected in how each encounter is documented: code **G10** (Huntington's disease) first, then layer the severity-specific neurocognitive disorder code: **F02.80**/**F02.81** for major NCD (without/with behavioral disturbance) or **G31.84 alone** for mild NCD, per DSM-5-TR coding rules.[4] Because psychiatric symptoms in HD are **physiologically driven** by the disease itself, they must be coded with **F06.x** etiological codes (e.g., **F06.31** depressive disorder due to HD, **F06.4** anxiety disorder due to HD), **not** primary psychiatric codes like F32.x or F41.x, which misrepresent the clinical picture and undercount disease complexity.*

*Each motor and somatic manifestation that a given patient experiences (including chorea (**R25.8**), dystonia (**G24.8**), dysphagia (**R13.12**), aspiration pneumonia (**J69.0**), gait unsteadiness (**R26.81**), or weight loss (**R63.4**)) should be documented individually, because each reflects **real clinical work** and a distinct dimension of that patient's disease burden. This layered, patient-specific coding tells the story of each individual's HD, captures the full scope of what primary care is managing, and supports **accurate HCC mapping** across multiple risk-adjustment dimensions. In a disease where every patient's combination of symptoms is unique, the coding should be equally individualized.*

4 TREATMENT AND REFERRAL QUICK GUIDE

HD cannot be cured or reversed: all treatment targets **symptom management** and **quality of life**.[16] Psychiatric symptoms (depression, anxiety, irritability) are **first-line treatment priorities** at any age, typically managed with SSRIs, though no HD-specific RCT evidence **guides agent selection**. **VMAT2 inhibitors** (tetrabenazine, deutetrabenazine, valbenazine) are reserved for functionally impairing chorea and require **heightened caution** in older LoHD patients, where **drug-induced parkinsonism**, **sedation**, and **dysphagia** compound age-related fall risk and polypharmacy burden. Tetrabenazine carries an FDA boxed warning for depression and suicidality. As chorea naturally diminishes in later stages, VMAT2 inhibitors should be **reassessed for deprescribing**, pivoting to **fall prevention** and **comfort care**. Pivotal VMAT2 trials enrolled predominantly younger, ambulatory patients, so **extrapolation to elderly LoHD** with comorbidities **requires caution**.

Primary care role: maintain suspicion for LoHD (especially without family history), screen suicidality **at every visit** (C-SSRS), monitor TFC and weight, coordinate the specialist team, and **integrate palliative care early** while the patient retains capacity for advance care planning. Palliative care in HD **is not end-of-life care**; it should begin at symptom onset, addressing **symptom burden**, **caregiver distress**, and advance directives **before** communication impairments limit patient participation

Treatment Pathway by Symptom

PRIORITY[16,39]	PREFERRED AGENT(S)	KEY GERIATRIC CAUTIONS
Depression/ suicidality (first-line; no RCT evidence)	**SSRIs**: sertraline 50–200 mg/day or citalopram 10–20 mg/day	Fluoxetine and paroxetine are strong **CYP2D6 inhibitors** = require VMAT2 inhibitor **dose reduction** if co-prescribed;*[39] C-SSRS at **every visit**
Irritability/agitation	**SSRI first**; low-dose atypical antipsychotics if inadequate (olanzapine 2.5–10 mg/day per 2025 Neuro-HD trial; **open label RCT**)	**Minimize** neuroleptic use in elderly: fall risk, sedation, EPS; **no RCT evidence** for HD psychiatric interventions
Apathy	Environmental structuring, activity scheduling = **first-line**; pharmacologic options limited	Distinguish **apathy from depression**: apathy responds poorly to SSRIs[16]
Chorea (functionally impairing)	**deutetrabenazine** (Austedo) 6 mg BID → max 48 mg/day (36 for CYP2D6 PMs); **valbenazine** (Ingrezza) 40 mg QD → max 80 mg/day = both FDA-approved[39]	Monitor depression, suicidality, parkinsonism, akathisia, QTc;*[39] **CONTRAINDICATED** in untreated depression or suicidality; Tetrabenazine **least preferred in elderly** (depression 23%, sedation 24%, parkinsonism 15%)*
Late-stage rigid-bradykinetic management	**DEPRESCRIBE VMAT2 inhibitors** as chorea diminishes; **dysphagia management**; nutritional support; polypharmacy review[39]	VMAT2 inhibitors **WORSEN** rigidity in **late HD**;[39] pivot from chorea suppression to **functional preservation** and **comfort care**

ABBREVIATIONS: RCT = randomized controlled trial; SSRI = selective serotonin reuptake inhibitor; CYP2D6 = cytochrome P450 2D6; VMAT2 = vesicular monoamine transporter 2; C-SSRS = Columbia Suicide Severity Rating Scale; EPS = extrapyramidal symptoms; HD = Huntington disease; PMs = poor metabolizers; FDA = US Food and Drug Administration; QTc = corrected QT interval; BID = twice daily; QD = once daily
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Non-Pharmacologic Care and Supportive Interventions

INTERVENTION	TARGET/RECOMMENDATION	NOTES
Dysphagia management (FEES + SLP)	**At symptom onset**; serial reassessment with progression[43]	**35%** early-stage, **94%** moderate, **100%** advanced dysphagia;[43] **aspiration pneumonia** 19.5% of HD deaths[11]

INTERVENTION	TARGET/RECOMMENDATION	NOTES
Modified diet textures (IDDSI)	Per **FEES findings**; thickened liquids when indicated[43]	**Reduces silent aspiration**; coordinate with caregivers and nursing home staff
Early PEG tube discussion	While patient retains **decision-making capacity**;[43] not as emergency response	HD-specific complexity: **gene-positive proxies** may have own decisional gaps[47]
Medical nutrition therapy	Energy expenditure **11–14% above** matched controls due to **hyperkinetic movements**;[44] individualize caloric targets using **adjusted Harris-Benedict equation**[48]	CPT **97802/97803**; track weight at **every visit**; code **R63.4** when weight loss documented; oral supplements (~500 kcal/day added) stabilized weight in 69% of patients[44]
Fall prevention: PT/OT	**Annual fall risk**; home safety review	Chorea, dystonia, gait instability all elevate fall risk; VMAT2 inhibitors **compound risk**[39]
Genetic counseling	Pre-test **and** post-test; family implications for **at-risk offspring** and proxies	ACMG 2014/2021 standards;[13,14] document **conflicts of interest** when proxy is at-risk
Early palliative care	Throughout disease course, **not just end-of-life**[47]	Advance care planning **while capacity intact**; CPT 99497/99498
Caregiver support and respite	HD **caregiver collapse** drives avoidable hospitalization[14]	**VBC priority**: integrate caregiver assessment; respite resources; HDSA/community support

ABBREVIATIONS: FEES = fiberoptic endoscopic evaluation of swallowing; SLP = speech-language pathology; HD = Huntington disease; IDDSI = International Dysphagia Diet Standardisation Initiative; PEG = percutaneous endoscopic gastrostomy; CPT = Current Procedural Terminology; PT = physical therapy; OT = occupational therapy; VMAT2 = vesicular monoamine transporter 2; ACMG = American College of Medical Genetics and Genomics; HDSA = Huntington's Disease Society of America; VBC = value-based care

Medication Safety and Key Interactions

DRUG/CLASS	INTERACTION/ TOXICITY*[3,9,10,16,25]	CLINICAL ACTION*[3,9,10,16,25]
deutetrabenazine (Austedo)	Depression, suicidality, parkinsonism, akathisia, QTc prolongation; CYP2D6 metabolism	C-SSRS at **every visit**; baseline + serial QTc; max 36 mg/day in CYP2D6 poor metabolizers;*[39] **CONTRAINDICATED** in untreated depression or suicidality

DRUG/CLASS	INTERACTION/ TOXICITY*[3,9,10,16,25]	CLINICAL ACTION*[3,9,10,16,25]
valbenazine (Ingrezza)	Depression, suicidality, parkinsonism, akathisia, QTc prolongation	C-SSRS at every visit; QTc monitoring; CONTRAINDICATED in untreated depression or suicidality[12]
tetrabenazine (least preferred in elderly)	Depression 23%, sedation 24%, parkinsonism 15% in pivotal RCT[39]	**Avoid** when deutetrabenazine or valbenazine accessible[39]
SSRIs (sertraline, citalopram)	May cause **initial symptom worsening**; CYP2D6-inhibiting SSRIs (fluoxetine, paroxetine) require VMAT2 inhibitor **dose reduction**	**Prefer sertraline or citalopram**; avoid fluoxetine and paroxetine if VMAT2 inhibitor co-prescribed[16]
Atypical antipsychotics (olanzapine, quetiapine)	Fall risk, sedation, extrapyramidal symptoms; metabolic effects	**Minimize in elderly**; use lowest effective dose; monitor metabolic labs and falls
Anticholinergics (oxybutynin, diphenhydramine, TCAs)	**Mask** early HD cognitive symptoms; independent dementia risk (OR 1.49)	Medication reconciliation at every visit; **deprescribe** per Beers Criteria[29]
Benzodiazepines	Sedation, falls, paradoxical agitation; Beers Criteria caution	**Avoid** except for acute agitation; coordinate with psychiatry
pyridostigmine, neuromuscular blockers	Aspiration pneumonia risk **amplifies with respiratory weakness** in advanced HD[11,16]	Document **dysphagia precautions**; SLP coordination
Dopamine-blocking antiemetics (metoclopramide, prochlorperazine)	**Can induce:** drug-induced parkinsonism and tardive dyskinesia (which mimics HD)[38]	**Avoid in HD** where alternatives exist; ondansetron or ondansetron + dexamethasone preferred

ABBREVIATIONS: QTc = corrected QT interval; CYP2D6 = cytochrome P450 2D6; C-SSRS = Columbia Suicide Severity Rating Scale; VMAT2 = vesicular monoamine transporter 2; RCT = randomized controlled trial; SSRI = selective serotonin reuptake inhibitor; HD = Huntington disease; TCA = tricyclic antidepressant; OR = odds ratio; SLP = speech-language pathology; FDA = US Food and Drug Administration

***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

When to Refer

CRITERION	SPECIALIST	URGENCY[16,18,19]
Active suicidal ideation; NMS; aspiration with respiratory distress	Emergency department	**EMERGENT**: C-SSRS; **discontinue** offending agent (NMS); airway management; code **J69.0** for aspiration
Confirmed or strongly suspected HD diagnosis	HD-specialist neurology	**URGENT** (within 1–2 weeks): **UHDRS motor exam**; TFC staging; CAG-repeat confirmation **if not yet tested**; coordinate long-term co-management plan
Genetic testing consideration	Genetic counseling (pre- and post-test)	**ROUTINE** (before test ordered): pre-test counseling required; informed consent; psychosocial readiness assessment; post-test counseling regardless of result
Refractory psychiatric symptoms; new psychosis	Psychiatry, preferably HD-experienced	**URGENT** (within 1–2 weeks): medication optimization; low-dose atypical antipsychotic consideration; distinguish HD-driven psychosis from delirium or polypharmacy effect
Dysphagia symptoms or weight loss	Speech-language pathology + FEES	**ROUTINE**, but **proactive** at any stage: **35%** early-stage dysphagia prevalence; serial reassessment with progression; **IDDSI diet modification** per findings; coordinate with caregivers
Falls, gait instability, functional decline	PT/OT + geriatric oncology/CGA	**ROUTINE** (2–4 weeks) if stable; **EXPEDITED** (1 week) if recent fall, hospitalization, or rapid TFC decline (≥2 points in 6 months)
Advance care planning while capacity intact	Palliative care	**ROUTINE**, but **early integration**: goals-of-care discussion; proxy designation (document conflicts if proxy is gene-positive); PEG tube preferences; CPT 99497/99498
End-of-life trajectory, hospice eligible	Hospice	**WHEN GOALS ALIGN**: transition when curative-intent interventions **no longer match** patient/family goals; coordinate with HD specialist and palliative team

ABBREVIATIONS: NMS = neuroleptic malignant syndrome; C-SSRS = Columbia Suicide Severity Rating Scale; HD = Huntington disease; UHDRS = Unified Huntington's Disease Rating Scale; TFC = Total Functional Capacity; CAG = cytosine-adenine-guanine; FEES = fiberoptic endoscopic evaluation of swallowing; IDDSI = International Dysphagia Diet Standardisation Initiative; PT = physical therapy; OT = occupational therapy; CGA = comprehensive geriatric assessment; PEG = percutaneous endoscopic gastrostomy; CPT = Current Procedural Terminology; SLP = speech-language pathology

Follow-Up Timing

STAGE/CATEGORY	FREQUENCY	LABS/MONITORING[16,18,19]
Pre-manifest/gene-positive (Z15.89)	**Annually** by neurology; primary care every **6 months**	Baseline cognitive battery (**MoCA**), **UHDRS motor exam** annually; psychological support
Stage I (TFC 11–13)	Neurology every **6 months**; primary care **quarterly**	TFC, MoCA, PHQ-9 + C-SSRS **at every visit**; weight; advance care planning **while capacity intact**
Stage II (TFC 7–10)	Neurology every **3–6 months**; primary care **quarterly**	TFC, MoCA, PHQ-9, C-SSRS, QTc if on VMAT2 inhibitor; **FEES baseline**; medication reconciliation
Stage III (TFC 3–6)	Neurology every **3 months**; primary care **monthly**	**Serial FEES**; weight and nutritional status; CGA; caregiver assessment
Stage IV–V (TFC 0–2)	Frequent **symptom-based** visits; palliative care primary	**Dysphagia management**; comfort-focused care; hospice when goals align
On VMAT2 inhibitor at any stage	**Monthly** during titration; **quarterly** during maintenance	**C-SSRS at every visit**; QTc; depression screen; **consider deprescribing** as chorea diminishes in late HD

ABBREVIATIONS: MoCA = Montreal Cognitive Assessment; UHDRS = Unified Huntington's Disease Rating Scale; TFC = Total Functional Capacity; PHQ-9 = Patient Health Questionnaire-9; C-SSRS = Columbia Suicide Severity Rating Scale; QTc = corrected QT interval; VMAT2 = vesicular monoamine transporter 2; FEES = fiberoptic endoscopic evaluation of swallowing; CGA = comprehensive geriatric assessment; HD = Huntington disease

Comorbidity Management — Primary Care Role

COMORBIDITY	APPROACH	CAUTION
Depression (F06.31/ F06.32)	SSRI first-line (sertraline, citalopram); PHQ-9 + C-SSRS at every visit	Code with F06.x physiological etiology codes, **NOT F32.x**
Suicidality risk	C-SSRS **at every visit**; psychiatry referral if positive	**~5x** general-population suicide risk;[10,46] VMAT2 inhibitors **contraindicated** with untreated suicidality

COMORBIDITY	APPROACH	CAUTION
Dysphagia	FEES; modified diet; SLP; early PEG discussion	Aspiration pneumonia **19.5%** of HD deaths[11]
Fall risk	Annual screening; PT/OT; home safety	VMAT2 inhibitors **compound fall risk** via parkinsonism[39]
Polypharmacy/ anticholinergic burden	Medication reconciliation **at every visit**	Anticholinergics **mask cognitive symptoms** and worsen HD
Caregiver burden	Assess **caregiver health** and gene status; respite resources[47]	Gene-positive proxies **may have own decisional gaps** → proactive navigation required

ABBREVIATIONS: SSRI = selective serotonin reuptake inhibitor; PHQ-9 = Patient Health Questionnaire-9; C-SSRS = Columbia Suicide Severity Rating Scale; VMAT2 = vesicular monoamine transporter 2; FEES = fiberoptic endoscopic evaluation of swallowing; SLP = speech-language pathology; PEG = percutaneous endoscopic gastrostomy; HD = Huntington disease; PT = physical therapy; OT = occupational therapy

Cost-Smart Options

BRAND (EST. COST)[49,50]	GENERIC/ ALTERNATIVE (EST. COST)[49,50]	MO. SAVINGS	COST-SMART TIP[51,52]
deutetrabenazine (Austedo) ~$5,800/mo standardized total cost; median OOP $63/mo (HD)	valbenazine (Ingrezza) **~$6,400/mo** standardized total cost; median **OOP $72/mo** (HD)	Both high cost	Both brand only; **manufacturer copay assistance available**; Medicare Part D OOP capped at **$2,000/yr** (IRA 2025+); MPPP spreads costs to **~$167/mo**
tetrabenazine (generic) ~$420/ mo total cost; ~$14/mo OOP	Generic tetrabenazine: **~$5,040/yr** total cost vs. **~$70,000–$77,000/yr** for Austedo/Ingrezza	**~$5,400–$6,000/mo** vs. brand VMAT2 inhibitors	**Lowest-cost VMAT2 option** but least preferred in elderly: higher depression, sedation, parkinsonism rates and TID dosing; reserve when Austedo/ Ingrezza inaccessible
Brand SSRIs (e.g., Lexapro/ escitalopram brand)	**Generic SSRIs**: sertraline **~$5–$15/ mo**; citalopram **~$5–$15/mo**	**~$200–$300/mo** vs. brand	**Generics clinically equivalent**;[53] avoid fluoxetine/paroxetine if co-prescribing VMAT2 inhibitors (strong CYP2D6 inhibitors)

ABBREVIATIONS: HD = Huntington disease; OOP = out-of-pocket; IRA = Inflation Reduction Act; MPPP = Medicare Prescription Payment Plan; VMAT2 = vesicular monoamine transporter 2; TID = three times daily; SSRI = selective serotonin reuptake inhibitor; CYP2D6 = cytochrome P450 2D6

Patient Education and Adherence

What makes HD patient education unique is the **depth of caregiver involvement** required, and the reality that the **caregiver themselves** may be at genetic risk. HD is autosomal dominant with **50% offspring risk**, meaning a spouse-child caregiver or sibling proxy **may carry the same gene** and face their own disease trajectory.[36] This creates a layer of complexity rarely seen in other neurodegenerative diseases: **the person making care decisions** may need genetic counseling, psychosocial support, and eventually their own care plan. HD caregiver collapse, driven by the combined burden of behavioral management, progressive dependency, and personal genetic uncertainty, is a recognized driver of **avoidable hospitalization**. **The PCP role** extends beyond the patient to identifying caregiver strain early, connecting families with community resources (HDSA support groups, respite care), and coordinating genetic counseling for at-risk proxies. **Early palliative care** should be integrated throughout the HD trajectory (**not** reserved for end-of-life) with advance care planning conversations initiated **while the patient retains decision-making capacity**.

DOCUMENTATION — PATIENT EDUCATION

*Document the following education topics, the patient and caregiver(s) present, materials provided, and patient understanding (**teach-back method**):*

- *Diagnosis and progressive nature in plain language*
- *Advance care planning and proxy designation (**document conflicts** if proxy is gene-positive)*
- ***Suicidality awareness**: symptom recognition and emergency contacts*
- *Dysphagia warning signs and **safe-swallow strategies***
- *VMAT2 inhibitor monitoring (depression, suicidality, when to call)*
- ***Genetic implications** for offspring and family members*
- *Caregiver resources (HDSA), respite, and gene-status considerations*

*Document whether the caregiver is a **known or potential gene carrier**, as this affects proxy decision-making and may warrant independent genetic counseling referral. CPT 99497/99498 are billable for advance care planning during the AWV without patient copay. Thorough documentation reflects the **multidimensional complexity** of HD management and supports closed-loop multidisciplinary coordination.*

Quality Metrics Tie-In

No HD-specific HEDIS or MIPS measures currently exist, but HD intersects multiple quality measurement domains simultaneously: **dementia management** (cognitive assessment, severity staging), **behavioral health** (depression screening, suicide risk), **geriatric safety** (fall risk, high-risk medications, medication reconciliation), and **goals-of-care documentation** (advance care planning). The disease's triad of motor, cognitive, and psychiatric manifestations, combined with **polypharmacy risks** from VMAT2 inhibitors, antidepressants, and antipsychotics, means that cross-cutting national measures apply **across the entire care trajectory** and can anchor quality reporting for any PCP managing an HD patient in a value-based program. Accurate ICD-10 coding (G10 + F02.x with severity tier) is the prerequisite for mapping HD patients in dementia-related measure denominators.

MEASURE	STANDARD	APPLICABILITY TO HD
HEDIS C08: Care for Older Adults (COA)- Medication Review	**Denominator**: MA enrollees ≥66, continuously enrolled **Numerator**: 4 sub-measures documented annually: **(1)** functional status assessment, **(2)** medication review, **(3)** pain assessment, **(4)** advance care planning **Exclusions**: hospice, ESRD	Medication discrepancies occur in **37–43%** of HD patients regardless of disease stage; cognitive impairment is the strongest predictor of discrepancies — pharmacy verification recommended over self-report
HEDIS COA: Care for Older Adults- Functional Status Assessment	**Denominator**: enrollees ≥66 **Numerator**: functional status assessment documented annually **Exclusions**: hospice, ESRD	**TFC score** (Shoulson-Fahn staging) directly satisfies this sub-measure; document TFC at every annual visit to track functional trajectory
HEDIS DAE: Use of High-Risk Medications in Older Adults	**Denominator**: enrollees ≥66 with dementia (F02.x) **Numerator**: ≥2 fills of same high-risk medication during year (lower is better) **Exclusions**: hospice	Neuroleptics, benzodiazepines, and anticholinergics are all on Beers Criteria; review for **HD-appropriate use**: some antipsychotics may be clinically necessary for HD psychosis but require documented justification
HEDIS DDE: Drug-Disease Interactions in Dementia	**Denominator**: enrollees ≥66 **Numerator**: dispensing of antipsychotics, benzodiazepines, or other drugs that exacerbate dementia (lower is better)	Antipsychotic use **for HD chorea/ psychosis** requires documented clinical rationale to avoid flagging as inappropriate
HEDIS: Medication Reconciliation Post-Discharge (MRP)	**Denominator**: enrollees ≥18 discharged from inpatient **Numerator**: medication reconciliation within 30 days **Exclusions**: hospice	Aspiration pneumonia and falls are primary HD **readmission drivers**; reconciliation critical for **VMAT2 inhibitors** + antidepressant + antipsychotic polypharmacy post-hospitalization
MIPS #047: Advance Care Planning	**Denominator**: patients ≥65 with ≥1 qualifying encounter during reporting period **Numerator**: advance care plan or surrogate decision-maker documented, OR documentation that ACP was discussed but patient declined **Exclusions**: none specified	Initiate while **decisional capacity intact**; HD-specific complexity: family proxies may **also be gene-positive**; billable as CPT 99497/99498 during AWV
MIPS #130: Documentation of Current Medications	**Denominator**: all patients with encounters **Numerator**: current medications documented at each visit	HD polypharmacy (VMAT2 inhibitors, SSRIs, antipsychotics) requires reconciliation **at every encounter**; cognitive impairment impairs self-report accuracy

MEASURE	STANDARD	APPLICABILITY TO HD
MIPS #134: Depression Screening (PHQ-9)	**Denominator**: patients ≥12 with ≥1 qualifying encounter **Numerator**: screened for depression using standardized tool (PHQ-9) AND follow-up plan documented if positive **Exclusions**: active depression diagnosis with current treatment	HD-related depression affects **~65%** of gene carriers and carries **~5x** suicide risk;[10,11] PHQ-9 at every visit with C-SSRS if positive; document follow-up plan

ABBREVIATIONS: HEDIS = Healthcare Effectiveness Data and Information Set; COA = Care for Older Adults; MA = Medicare Advantage; ESRD = end-stage renal disease; HD = Huntington disease; TFC = Total Functional Capacity; VMAT2 = vesicular monoamine transporter 2; MIPS = Merit-based Incentive Payment System; ACP = advance care planning; CPT = Current Procedural Terminology; AWV = annual wellness visit; SSRI = selective serotonin reuptake inhibitor; PHQ-9 = Patient Health Questionnaire-9; C-SSRS = Columbia Suicide Severity Rating Scale; MoCA = Montreal Cognitive Assessment; MRP = Medication Reconciliation Post-Discharge

QUALITY OUTCOME

*When primary care recognizes HD pattern **early** (treatment-resistant depression + subtle motor signs + family history, even without prior HD diagnosis), supports **genetic counseling referral**, codes manifestations **comprehensively** (G10 + F02.x severity tier + F06.x psychiatric + R-codes for motor/somatic), screens suicidality **at every visit**, manages dysphagia **proactively**, and integrates palliative care **while capacity intact**, patients experience earlier diagnosis, fewer aspiration hospitalizations, lower fall-related ED visits, and meaningful advance care planning. LoHD accounts for **~11% of HD cases** and up to 31% **lack known family history**,[3,12] making **primary care pattern recognition** the critical diagnostic gateway. Quality outcomes follow naturally from delivering and documenting the care the clinical picture demands: HCC capture, quality composites, and care coordination metrics are **consequences of good care**, not separate goals.*

5 CODING REMINDERS AND CASE EXAMPLES

Documentation Specificity

- **G10 (Huntington's disease)**: Code **G10** at every HD encounter = confirmed by **CAG ≥36** on molecular genetic testing (CPT 81271) or **clinical motor DCL 4** with family history. Document CAG repeat length and TFC stage in the assessment
- **Major NCD due to HD**: Code **first G10**, **then** the severity-specific **F02.x code**:[4] **F02.A0–A4** (mild severity), **F02.B0–B4** (moderate), or **F02.C0–C4** (severe), with 5th/6th characters specifying accompanying behavioral disturbance, psychotic disturbance, mood disturbance, or anxiety. When more than one disturbance is present, **code each separately** (e.g., G10 +

F02.C11 + F02.C2 + F02.C3 for severe major NCD with agitation, psychotic disturbance, and mood symptoms)

- **Mild NCD due to HD**: Code **first G10**, **then F06.70** (without behavioral disturbance) or **F06.71** (with behavioral disturbance).[4] Do **NOT** use G31.84 for mild NCD due to HD → G31.84 is reserved for mild NCD due to **possible or unknown etiology** only
- **Psychiatric manifestations**: Use **F06.x** physiological-etiology codes to link psychiatric symptoms directly to HD:[4] **F06.31**/**F06.32** (depressive), **F06.4** (anxiety), **F06.0** (psychotic with hallucinations), **F06.2** (psychotic with delusions). Do **NOT** use primary psychiatric codes (F32.x/F41.x) → implies a primary psychiatric disorder unrelated to HD and **misses etiological linkage**[4]
- **Motor and somatic manifestations**: Document each **individually**: **R25.8** (chorea), **G24.8** (dystonia), **R13.12** (oropharyngeal dysphagia), **J69.0** (aspiration pneumonia), **R26.81** (unsteadiness/falls), **R63.4** (weight loss)
- **Family history/pre-manifest status**: **Z82.0** (family history of epilepsy and other diseases of the nervous system) for at-risk relatives; **Z15.89** (genetic susceptibility) for **gene-positive pre-manifest individuals** not yet meeting diagnostic criteria

Annual Clinical Review and Confirmation

- **Annual review: Active HD (G10)** requires **annual MEAT documentation**; manifestations **(F02.x, F06.x, R-codes)** refreshed at each encounter
- **Visit modality:** Face-to-face **or** synchronous audio-video telehealth qualifies when it supports meaningful evaluation
- **Clinical context: G10** maps to **HCC 200** (CNA RAF 0.279);[7,8] F02.x dementia by severity maps HCC 125/126/127 (all RAF 0.341);[7,8] both can map **concurrently with proper sequencing**
- **Avoid rollover: Do not** copy forward last year's HD note **without updating**: TFC stage, CAG status, psychiatric/motor/dysphagia progression, and advance care planning status

Good Documentation is Comprehensive Coding

EHR SHORTCUT/RISK	PREFERRED DOCUMENTATION LANGUAGE
Coding G10 alone	'Active HD (**G10**), **Stage II** per TFC 10; major NCD moderate severity (**F02.B0**); depression due to HD (**F06.31**); chorea (**R25.8**); CAG 43 confirmed [date]'
Using F32.x for HD-related depression	Use **F06.31**/**F06.32** (mood due to physiological condition) → links symptom to HD etiology[4]

EHR SHORTCUT/RISK	PREFERRED DOCUMENTATION LANGUAGE
G10 + G31.84 combination	**G31.84 alone** for mild NCD → do **NOT** add G10[7]
'On Austedo' without monitoring documentation	'Deutetrabenazine 36 mg/day for HD chorea; **monthly C-SSRS**, **depression screen**, QTc; not currently rigid-bradykinetic (TFC 10)'
'Dysphagia' (HD patient)	'Oropharyngeal dysphagia due to HD (R13.12 + G10); **FEES** [date] phase-specific findings; **IDDSI level 4 modified diet**; SLP managing'

ABBREVIATIONS: HD = Huntington disease; TFC = Total Functional Capacity; NCD = neurocognitive disorder; CAG = cytosine-adenine-guanine; C-SSRS = Columbia Suicide Severity Rating Scale; QTc = corrected QT interval; FEES = fiberoptic endoscopic evaluation of swallowing; IDDSI = International Dysphagia Diet Standardisation Initiative; SLP = speech-language pathology

EHR Workflow Tips

- **[EHR TIP — Smartphrase]** Build a .hd dot phrase that **auto-populates G10** + **severity-tier F02.x** (or G31.84 for mild NCD) + **F06.x** psychiatric codes + **R-codes** for motor manifestations, **TFC stage**, **CAG repeat** length, C-SSRS result, and medication review (anticholinergic/neuroleptic/VMAT2 inhibitor status) = sourced from neurology notes, genetic testing, and cognitive assessments. Reduces **G10-only coding** and supports MEAT documentation in a single paste
- **[EHR TIP — Alert]** No neurology visit **in 12 months**: configure a **gap alert** for any active G10 without a documented HD-specialist neurology encounter in the past 12 months. Surfaces patients who may have fallen out of multidisciplinary follow-up during care transitions or hospitalizations
- **[EHR TIP — Alert] No suicide risk screening on VMAT2 inhibitor:** fire a secondary alert for active G10 + VMAT2 inhibitor prescription **without C-SSRS documented in the past 90 days**. VMAT2 inhibitors carry an FDA boxed warning for depression and suicidality; HD independently confers **2–9x** suicide risk
- **[EHR TIP — Alert] No advance care planning documented**: fire an alert for active G10 without ACP (CPT 99497) on file. Supports **MIPS #047** compliance and early goals-of-care documentation while decisional capacity remains intact
- **[EHR TIP — Best Practice] Problem list hygiene:** maintain active **G10** with severity-specific **F02.x** (**not** unspecified F02.80), each psychiatric manifestation coded **individually** (F06.31 depression, F06.4 anxiety), **motor manifestation**s (R25.8 chorea, R26.81 gait instability, R13.12 dysphagia), **CAG repeat length**, and current **TFC stage**. DSM-5-TR requires G10 **sequenced first** when paired with F02.x;[4] G31.84 **stands alone** without G10 for mild NCD[4]
- **[EHR TIP — Order Set] HD initial workup bundle**: HTT CAG-repeat genetic test (CPT 81271), MoCA, PHQ-9, C-SSRS, MRI brain, TSH, B12, comprehensive metabolic panel. **New-**

diagnosis add-on bundle includes HD-specialist neurology referral, genetic counseling, psychiatry consult, and SLP baseline evaluation → all triggered at **G10 problem list entry**

Brief Case Examples

SUCCESS: COMPREHENSIVE DOCUMENTATION

SCENARIO

67-year-old patient with **treatment-resistant depression** × 4 years and recent gait instability. Family history reveals a paternal aunt with "**early dementia**." PCP recognized the late-onset HD pattern, referred to genetic counseling, and ordered HTT testing: **CAG 43** repeats confirmed full penetrance.

Documentation: "Active Huntington disease (**G10**), late-onset, Stage II per Shoulson-Fahn **TFC 10**; CAG 43 repeats confirmed [date]. Major **NCD due to HD**, mild: **F02.A0** (coded **after G10** per DSM-5-TR). Depressive disorder due to HD, F06.31 (**NOT** primary F32.x). Chorea (**R25.8**). Sertraline 100 mg/day continued; C-SSRS positive ideation no plan → psychiatry consult placed. SLP referral for proactive dysphagia baseline (FEES: **no aspiration**); PT/OT for fall prevention; HD-specialist neurology managing; genetic counseling addressing family implications; advance care planning initiated **while capacity intact** (CPT 99497)."

Outcome: **HCC 200** (G10) and **HCC 127** (F02.A0) both mapped per **DSM-5-TR-correct sequencing**;[4,7,8] psychiatric symptoms coded with **physiological etiology** (F06.x) preserving HD linkage; proactive dysphagia and fall management documented; advance care planning supports COA composite.

PITFALL: INSUFFICIENT DOCUMENTATION

Documentation as written: "Huntington's disease." Coded as **G10 alone**. The chart references "**moderate dementia**" in a neurology note from 6 months ago with behavioral disturbance; patient is on **sertraline** for depression; chorea managed with deutetrabenazine; recent FEES showed pharyngeal-phase dysphagia. **None of these manifestations** appears on the active problem list; only G10.

Consequence: G10 alone **misses HCC 127/126/125** (dementia, RAF 0.341); the absence of F02.x severity coding fails the DSM-5-TR sequencing rule;[4] depression coded (if at all) as F32.x rather than F06.31 **obscures HD etiology**; chorea (R25.8) and oropharyngeal dysphagia (R13.12) not on problem list, missing clinical-complexity documentation. VMAT2 inhibitor monitoring (C-SSRS, depression, QTc) **not documented** despite FDA label requirements.

RAF Impact: Lost **HCC 126** mapping for moderate F02.B dementia (**CNA RAF 0.341** ≈ $3,547/year illustrative); missed comorbidity coding for chorea, dysphagia, and HD-attributed depression **understates the actual clinical complexity** the team is managing.

Fix: Update problem list and assessment: "Active Huntington disease (**G10**): **Stage III** per TFC 5; major NCD due to HD moderate with behavioral disturbance = **F02.B11** (DSM-5-TR sequence); mood disorder due to HD with major depressive-like episode: **F06.32** (**NOT** F32.x); chorea: **R25.8**; oropharyngeal dysphagia: **R13.12** (FEES [date]). **CAG 44** confirmed; on deutetrabenazine 36 mg/day with **monthly C-SSRS**, depression screen, and QTc." Document MEAT **for each** at the next encounter; place SLP referral if not already in place.

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