



Managing Aggression in Huntington's Disease

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Presenter Disclosures

Garima Arora, MD

The following personal financial relationships with commercial interests relevant to this presentation existed during the past 12 months:

**No relationships to disclose
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**Huntington's Disease
Society of America**

Aggression in Huntington's Disease

- ❖ Very common (22 to 66%) in Huntington's Disease
- ❖ Impacts patient, caregivers, and care-facilities
- ❖ More common in males
- ❖ More commonly during early stages of disease
 - ✓ can even occur prior to onset of motor symptoms
 - ✓ mostly 6 to 11 years after onset of HD

Aggression in Huntington's Disease

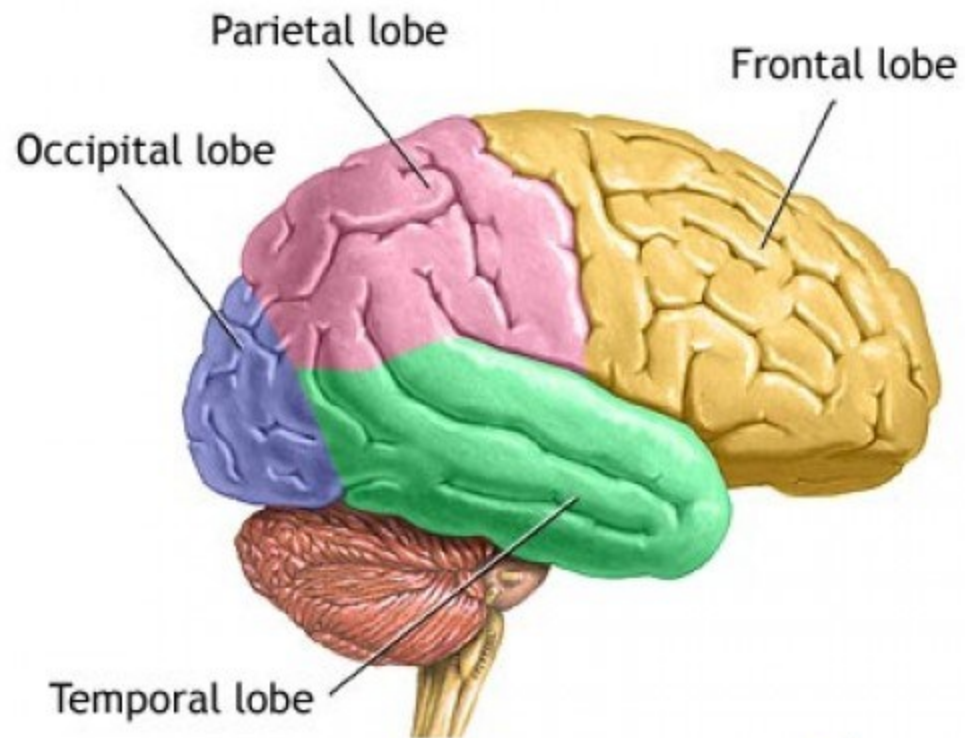
- ❖ One of the primary causes of hospitalization
- ❖ May lead to suicide or assault or homicide
- ❖ May have criminal and legal consequences

Aggression in Huntington's Disease

- ❖ from Latin *aggredi* – to attack
- ❖ Is a pattern of behavior directly observed by family members, acquaintances, caregivers, and/or providers
- ❖ Not the same as irritability which is a mood state

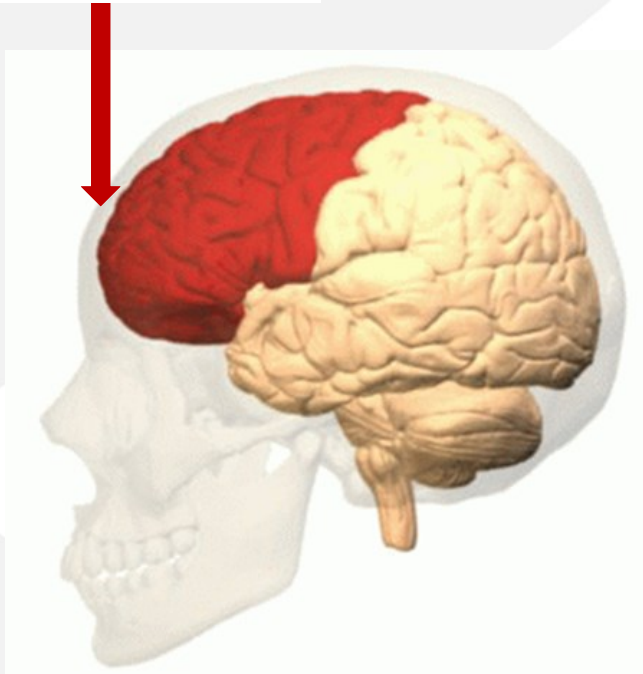
Aggression in Huntington's Disease

- ❖ **Physical Aggression:** Behavior manifested with intent to inflict physical harm or damage to a person or animate/ inanimate object
- ❖ **Verbal Aggression:** Behavior manifested with intent to threaten or intimidate or reject the recipient

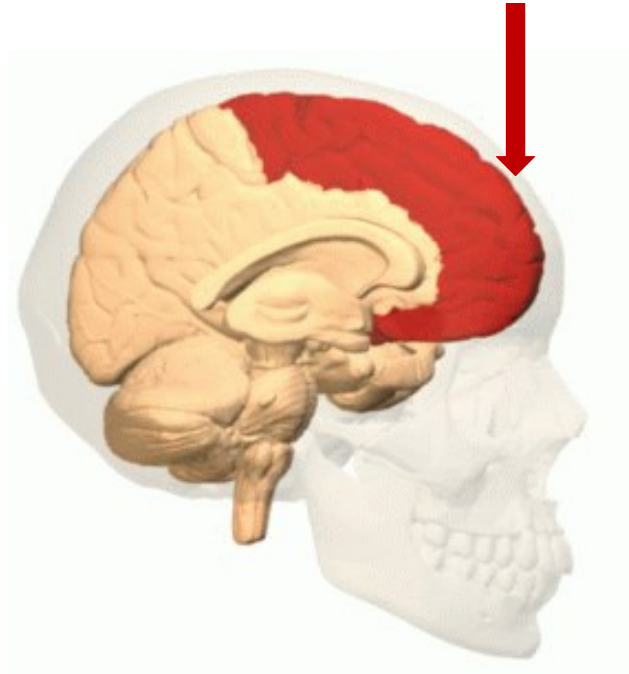


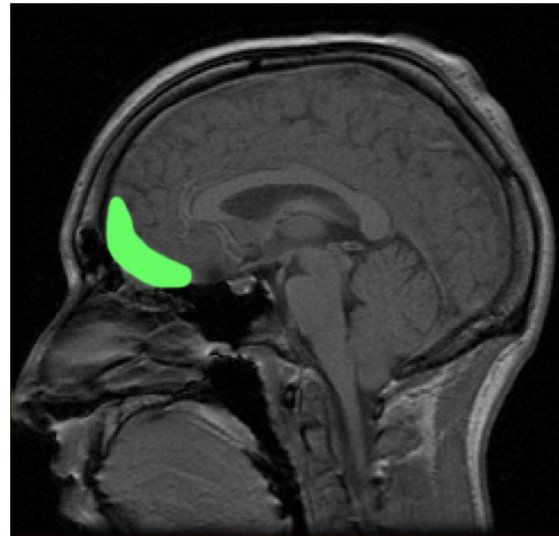
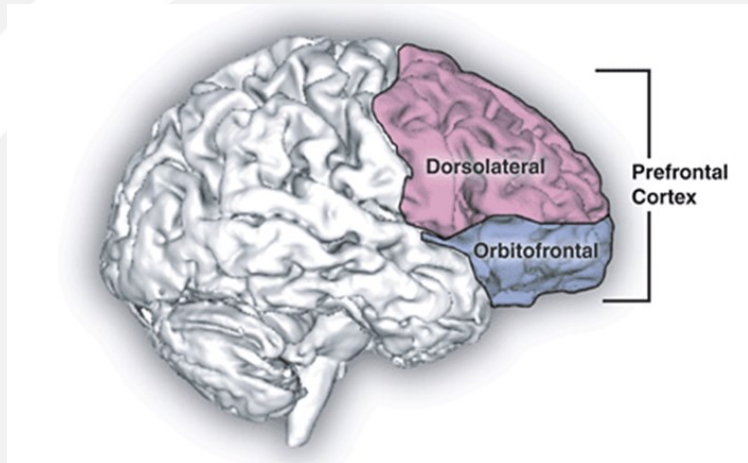
ADAM.

Prefrontal Cortex



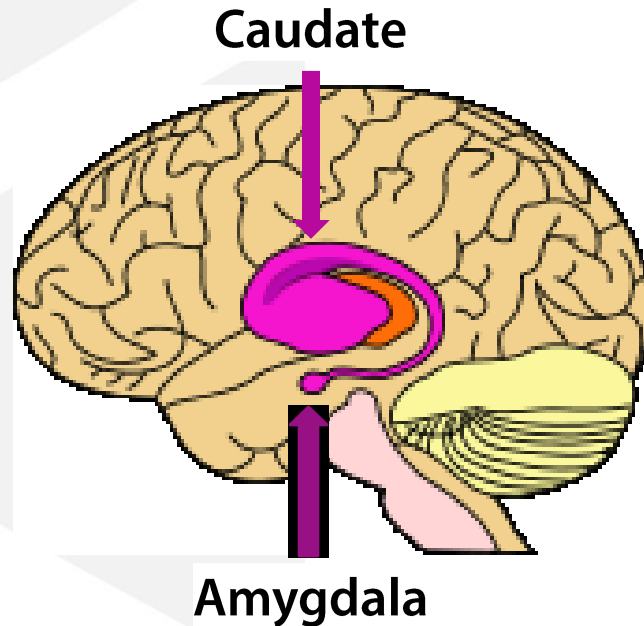
Prefrontal Cortex



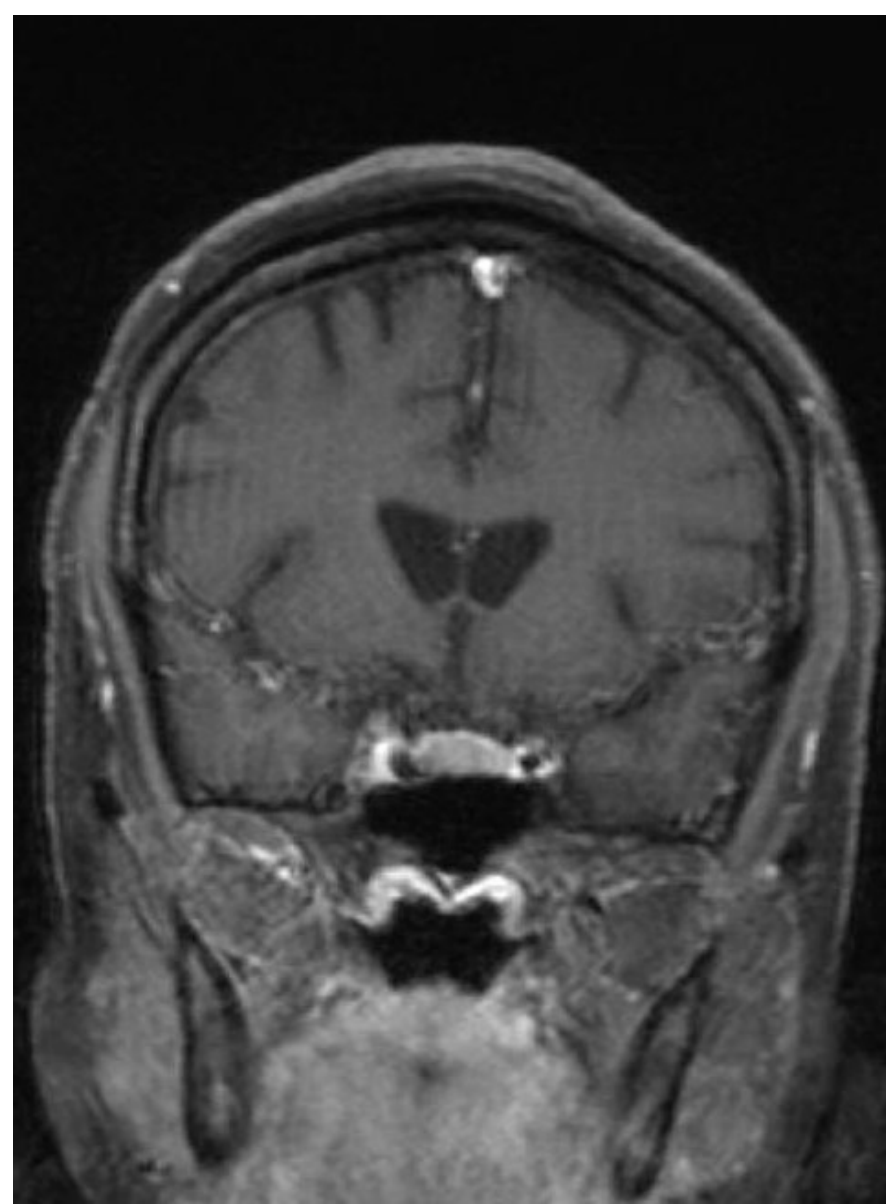
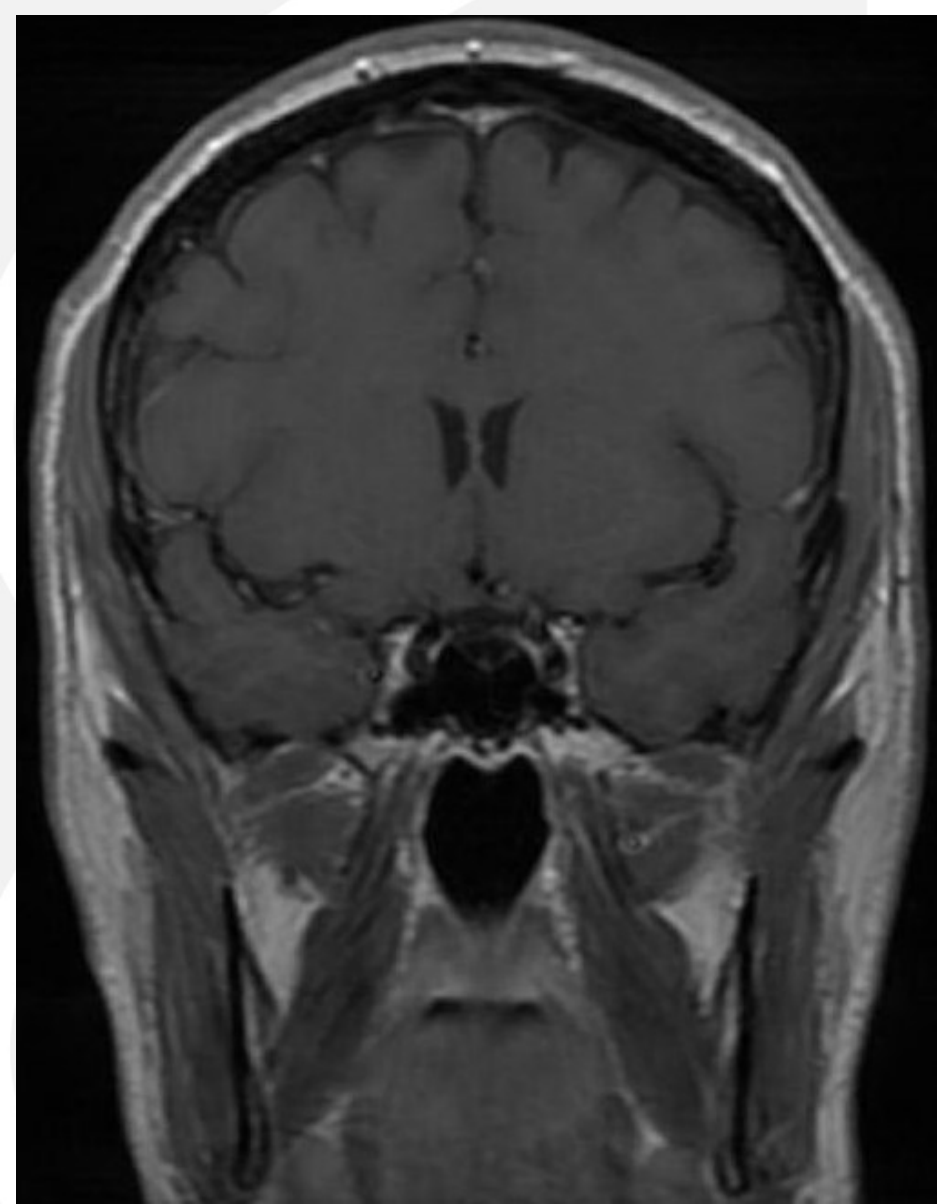


OFC plays a key role in impulse control, monitoring ongoing behavior and socially appropriate behavior

- **Relay Station between Limbic System and Frontal Lobes**
- **Damaged in Huntington's**



- **Associated with forming emotional responses, particularly involving aggression**



Aggression in Huntington's Disease

- ❖ Quick shifts in behavior
- ❖ Unpredictable responses to stimuli
- ❖ *More Intense* response to triggers
- ❖ *Explosive* emotional outburst
- ❖ Individual unaware of behavior and/ or intensity
- ❖ Decreased insight with progression of disease

Aggression in Huntington's Disease

Exemplars of agitated/ aggressive behavior

- ❖ Not-redirectable *temper tantrum*
- ❖ Yelling incessantly
- ❖ Constant profanity or foul language
- ❖ Threatening statements
- ❖ Throwing things

Aggression in Huntington's Disease

Exemplars of agitated/ aggressive behavior

- ❖ Punching objects/ walls/ doors
- ❖ Banging head against the wall
- ❖ Threatening gestures
- ❖ Physical violence

Aggression in Huntington's Disease

Can be indicative or manifestation of other emotions

❖ Hunger/ Thirst

❖ Pain

❖ Discomfort

❖ Fear

Aggression in Huntington's Disease

Can be indicative or manifestation of other emotions

❖ Frustration

- Inability to Communicate
- Loss of ability to do a task

❖ Depression/ Anxiety

❖ Delusions/ Hallucinations

Aggression in Huntington's Disease

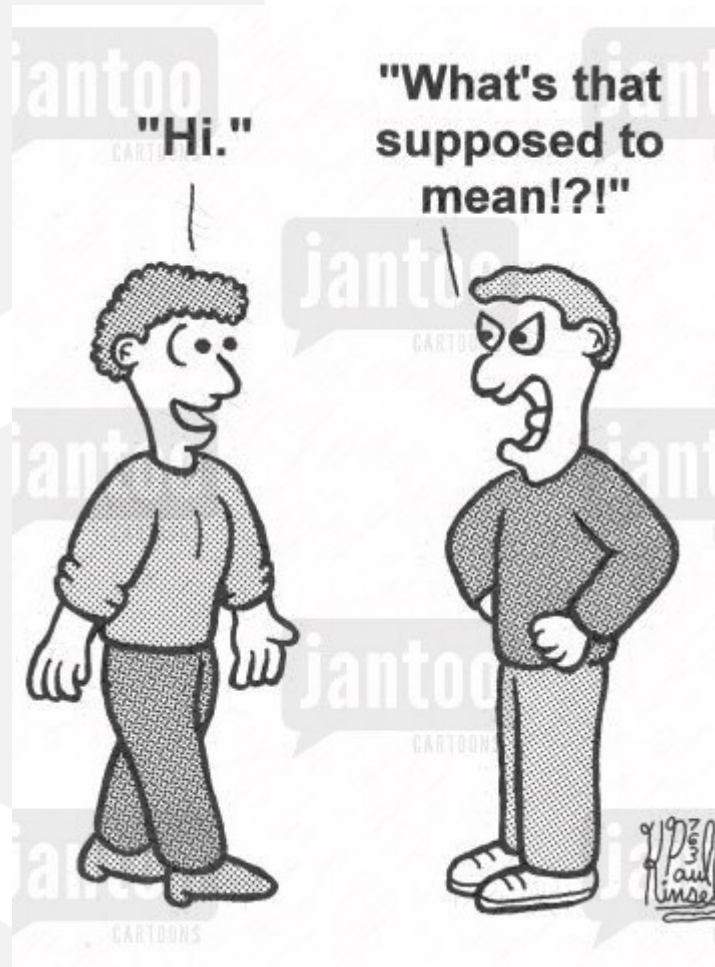
Can also be secondary to:

- ❖ Electrolyte abnormalities
- ❖ Hypoglycemia/ Hyperglycemia
- ❖ Infections, particularly UTIs
- ❖ Medication side-effects
- ❖ Metabolic Dysfunction
- ❖ Trauma
- ❖ Stroke, Shock, MI, PE
- ❖ **I WATCH DEATH**

Aggression in Huntington's Disease

Common Triggers:

- ❖ Fatigue
- ❖ Poor sleep
- ❖ Change in routine
- ❖ New caregiver or provider
- ❖ Failure to get their own way
- ❖ Challenges to perseverating thoughts or delusions
- ❖ Perceived or real mistreatment



Aggression in Huntington's Disease

Assessment and Diagnosis:

- ❖ Observed reports
- ❖ Clinical Interview
- ❖ Rating Scales
 - ❖ UHDRS (Unified Huntington's Disease Rating Scale)
 - ❖ PBA-HD (Problem Behaviors Assessment for Huntington's Disease)
 - ❖ RAGE (Rating Scale for Aggressive Behavior in the Elderly)
 - ❖ Yudofsky Aggression Scale
 - ❖ Cohen-Mansfield Agitation Inventory (CAMI)
 - ❖ Neuropsychiatric Inventory (NPI) – agitation scale

Aggression in Huntington's Disease

Good News

- ❖ Symptoms responsive to treatment
- ❖ Multiple treatments : pharmacologic and non-pharmacologic
- ❖ Treatments can help control both the frequency and intensity of aggressive behaviors

Aggression in Huntington's Disease

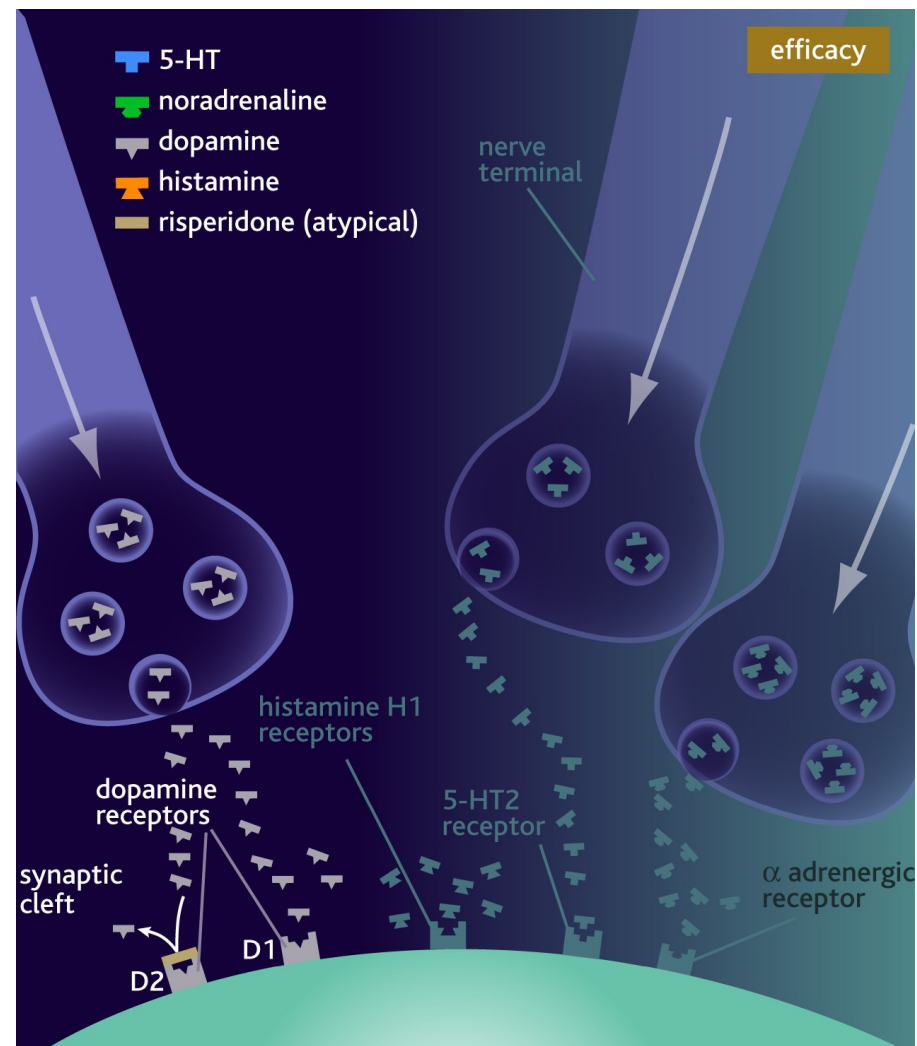
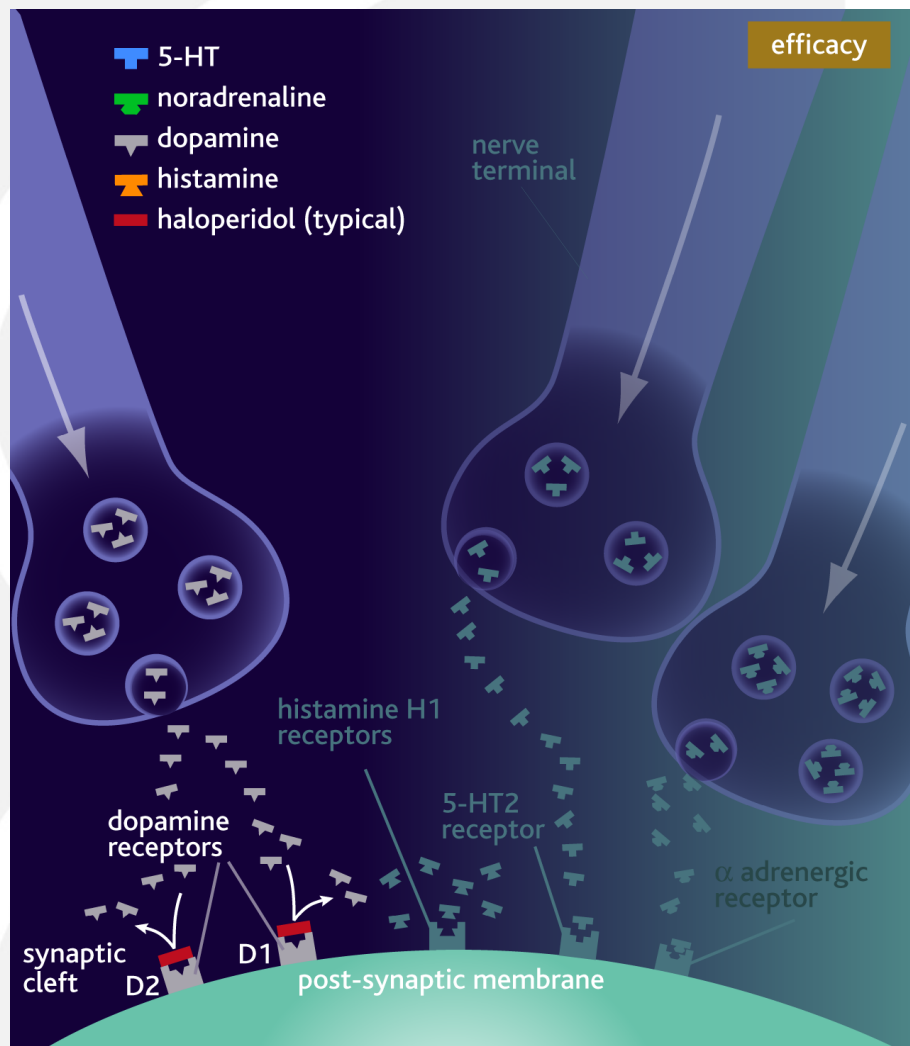
MEDICATIONS:

- ❖ ANTI-PSYCHOTICS
- ❖ ANTI-DEPRESSANTS
- ❖ ANTI-ANXIETY
- ❖ MOOD STABILIZERS
- ❖ OTHERS

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MEDICATIONS:

❖ ANTI-PSYCHOTICS



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ANTI-PSYCHOTICS:

❖ TYPICAL

- Clozapine (Clozaril), Haloperidol (Haldol), Thioridazine (Mellaril)

❖ ATYPICAL

- Aripiprazole (Abilify), Olanzapine (Zyprexa), Quetiapine (Seroquel), Risperidone (Risperdal)

Aggression in Huntington's Disease

ANTI-PSYCHOTICS:

Pros	Cons
<ul style="list-style-type: none">• Suppression of choreic movements• May help with underlying depression/ apathy	<ul style="list-style-type: none">• Sedation• Metabolic Syndrome

Aggression in Huntington's Disease

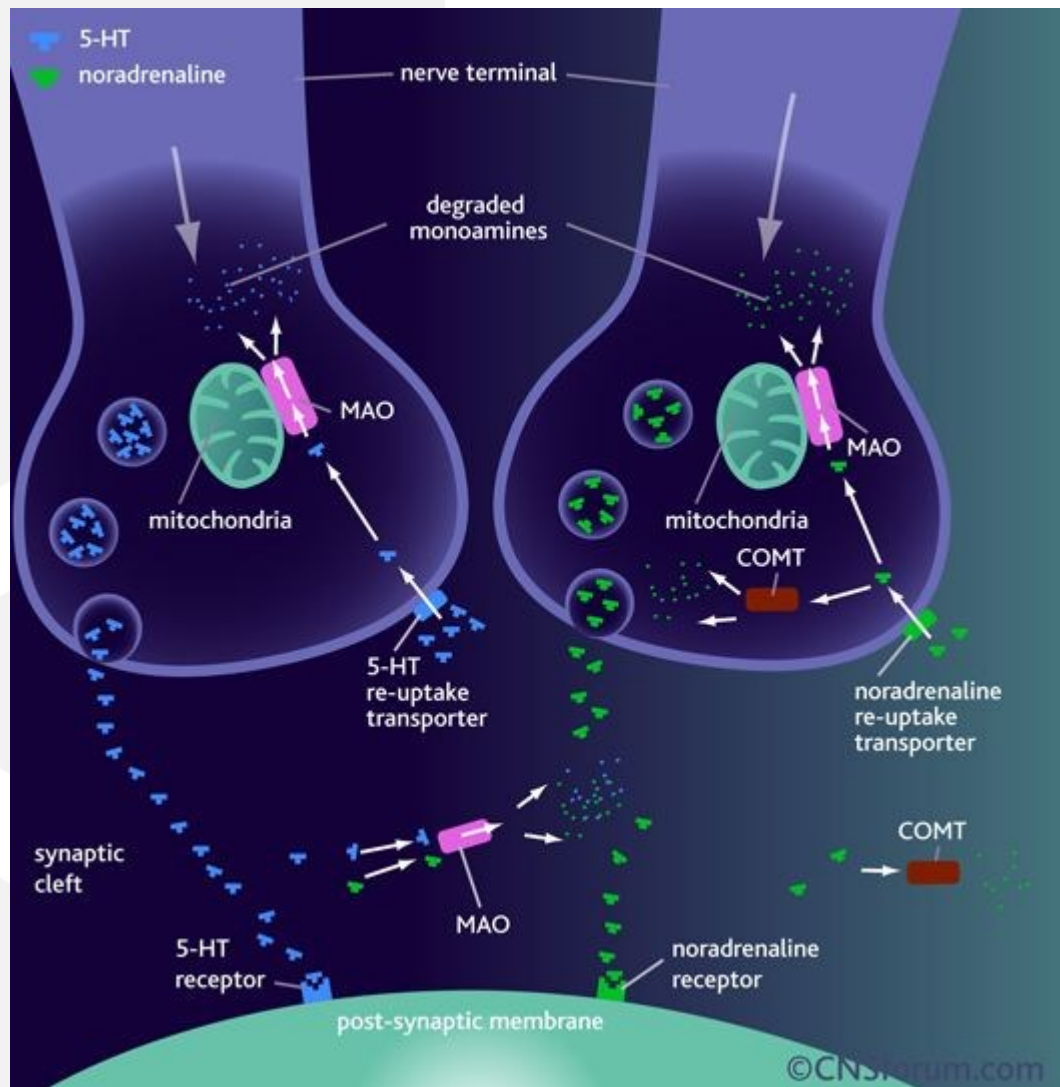
MEDICATIONS:

❖ ANTI-DEPRESSANTS

Aggression in Huntington's Disease

ANTI-DEPRESSANTS:

- ❖ SSRIs (Selective Serotonin Reuptake Inhibitors)
- ❖ SNRIs (Serotonin and Norepinephrine Reuptake Inhibitors)
- ❖ Norepinephrine Serotonin modulators
- ❖ Serotonin modulators



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Aggression in Huntington's Disease

ANTI-DEPRESSANTS:

- ❖ **SSRIs (Selective Serotonin Reuptake Inhibitors)**
 - Citalopram (Celexa), Escitalopram (Lexapro), Fluoxetine (Prozac), Paroxetine (Paxil), Sertraline (Zoloft), Fluvoxamine (Luvox)

Aggression in Huntington's Disease

ANTI-DEPRESSANTS:

- ❖ **SNRIs (Serotonin and Norepinephrine Reuptake Inhibitors)**
 - **Duloxetine (Cymbalta), Venlafaxine (Effexor),
Desvenlafaxine (Pristiq), Levomilnacipran (Fetzima)**

Aggression in Huntington's Disease

ANTI-DEPRESSANTS:

❖ Norepinephrine Serotonin modulators

➤ Mirtazapine (Remeron)

Aggression in Huntington's Disease

ANTI-DEPRESSANTS:

❖ Serotonin modulators

➤ Trazodone (Desyrel)

Aggression in Huntington's Disease

ANTI-DEPRESSANTS:

Pros	Cons
<ul style="list-style-type: none">• Treatment of underlying Depression or Anxiety or OCD	<ul style="list-style-type: none">• GI Distress• Headache

Aggression in Huntington's Disease

MEDICATIONS:

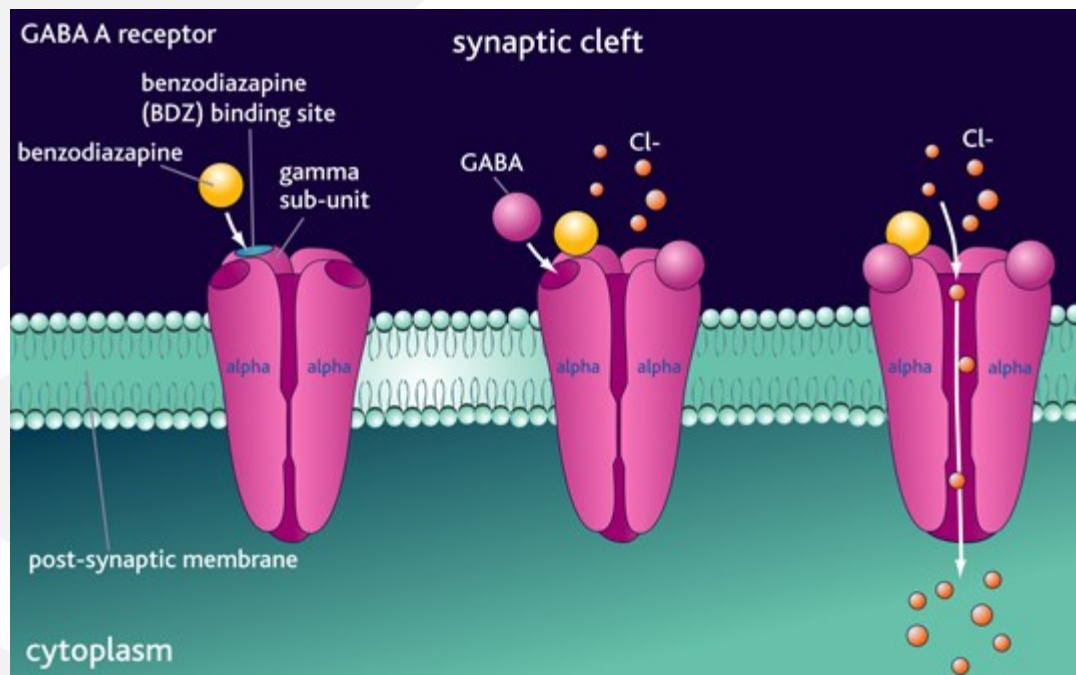
❖ **ANTI-ANXIETY**

Aggression in Huntington's Disease

ANTI-ANXIETY:

❖ BENZODIAZEPINES

- Alprazolam (Xanax), Clonazepam (Klonopin), Diazepam (Valium), Lorazepam (Ativan)



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Aggression in Huntington's Disease

BENZODIAZEPINES:

Pros	Cons
<ul style="list-style-type: none">• Treatment of underlying Anxiety or OCD• Assistance with Sleep	<ul style="list-style-type: none">• Disinhibition (may make agitation worse)• Confusion

Aggression in Huntington's Disease

ANTI-ANXIETY:

❖ BUSPIRONE (BUSPAR)

- Acts on Serotonin receptor (5HT-1A), Dopamine receptor, and Adrenergic receptors

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MEDICATIONS:

❖ MOOD STABILIZERS

Aggression in Huntington's Disease

MOOD STABILIZERS:

- ❖ LITHIUM

- ❖ ANTI-SEIZURE MEDICATIONS

 - Carbamazepine (Tegretol), Divalproex (Depakote),
Oxcarbazepine (Trileptal)

Aggression in Huntington's Disease

MOOD STABILIZERS:

Pros	Cons
<ul style="list-style-type: none">• May help with underlying depression/apathy• Divalproex helps with myoclonic hyperkinesia• Lithium may help with motor function• Lithium may reduce suicidality	<ul style="list-style-type: none">• Toxicity

Aggression in Huntington's Disease

MEDICATIONS:

❖ OTHERS

Aggression in Huntington's Disease

OTHERS:

- ❖ **BETA BLOCKERS**

 - **PROPRANOLOL**

- ❖ **AMANTADINE**

Aggression in Huntington's Disease

GENERAL CONSIDERATIONS ABOUT MEDICATIONS:

- ❖ Antipsychotics most commonly prescribed
- ❖ Efficacy fairly similar among different classes
- ❖ Variable response among patients
- ❖ Variable response in the same patient
- ❖ *Start low go slow*
- ❖ Scheduled vs. PRN

Aggression in Huntington's Disease

NON PHARMACOLOGIC TREATMENTS:

- ❖ EDUCATION
- ❖ BEHAVIORAL MODIFICATIONS
- ❖ COUNSELLING
- ❖ PREVENTING CAREGIVER BURN-OUT

Aggression in Huntington's Disease

STRATEGIES FOR BEHAVIORAL MODIFICATION

- ✓ *****SAFETY IS PRIMARY!!!*****
- ✓ Identify triggers and cues
- ✓ “Pick your battles”
- ✓ Keep calm
- ✓ Remember that patient has little or no insight

Aggression in Huntington's Disease

STRATEGIES FOR BEHAVIORAL MODIFICATION

- ✓ Confirm it is agitation and not other emotion
- ✓ Acknowledge the feeling behind the behavior
- ✓ Redirect or distract
- ✓ Time out
- ✓ Avoid confrontation or ultimatums
- ✓ Use simple and clear communications

Aggression in Huntington's Disease

STRATEGIES FOR BEHAVIORAL MODIFICATION

- ✓ Accept apologizes without recrimination
- ✓ Do not hesitate to seek additional help
- ✓ Discuss with your care providers
- ✓ *****ALWAYS BE AWARE!!*****

Aggression in Huntington's Disease

BEHAVIORAL MODIFIERS

- ✓ **Decrease caffeine and sugar intake**
- ✓ **Maintain structure and routine**
- ✓ **Maintain safe and calm environment**
- ✓ **Encourage to do activities within functioning capacity**

Aggression in Huntington's Disease

BEHAVIORAL MODIFIERS

- ✓ Music, Meditation, Reading
- ✓ Sensory modulation interventions
- ✓ Keep dangerous objects out of easy reach

Aggression in Huntington's Disease

BEHAVIORAL PLANNING (FACILITIES)

- ✓ **Person-centered care**
- ✓ **Specialized training for staff**
- ✓ **Maintain safe and calm environment**
- ✓ **Maintain structure and routine**

Aggression in Huntington's Disease

BEHAVIORAL PLANNING (FACILITIES)

- ✓ **Organized activities**
- ✓ **Sensory modulation interventions**
- ✓ **Mapping of care**

Resources

- **HOPES** (Huntington's Outreach Project for Education at Stanford):

http://web.stanford.edu/group/hopes/cgi-bin/hopes_test/

- **Huntington's Disease Society of America**

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/>

- <https://www.cnsforum.com/educationalresources>

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