



# Managing Aggression in Huntington's Disease

Garima Arora, MD



Huntington's Disease  
Society of America

The information provided by speakers in workshops, forums, sharing/networking sessions and any other educational presentation made as part of the 2015 HDSA Convention program is for informational use only.

HDSA encourages all attendees to consult with their primary care provider, neurologist or other healthcare provider about any advice, exercise, medication, treatment, nutritional supplement or regimen that may have been mentioned as part of any presentation.

# 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  
or list**



**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

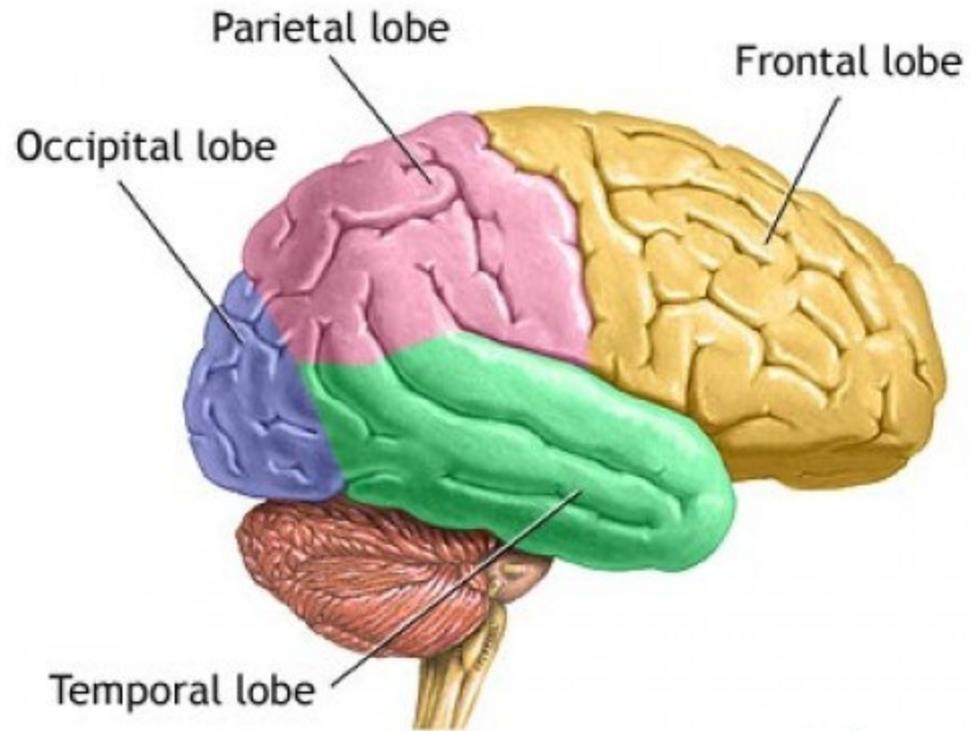
- ❖ One of the primary causes of hospitalization
- ❖ May lead to suicide or assault or homicide
- ❖ May have criminal and legal consequences

# 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

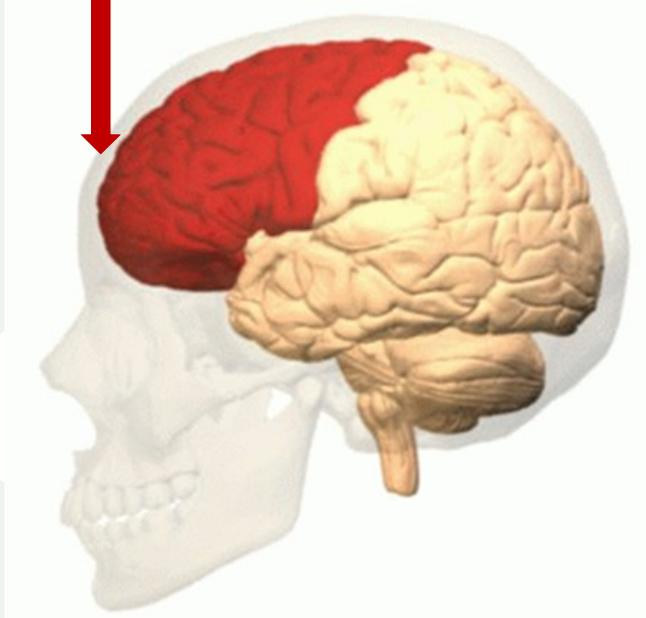
# Aggression in Huntington's Disease

- ❖ **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

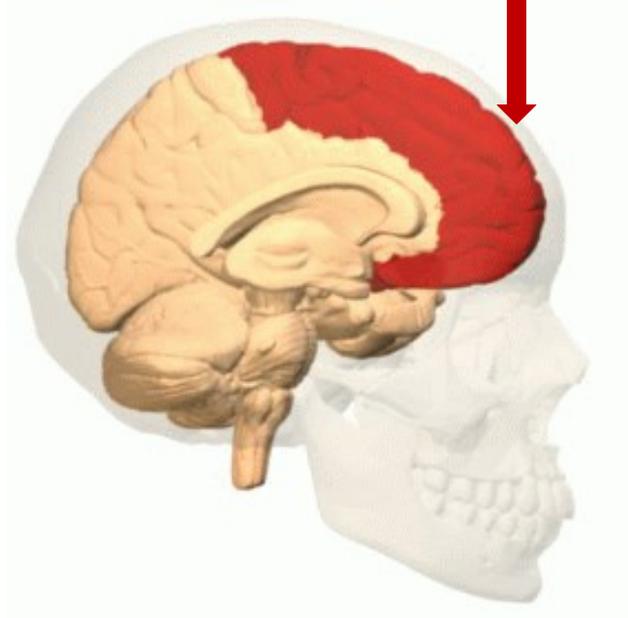


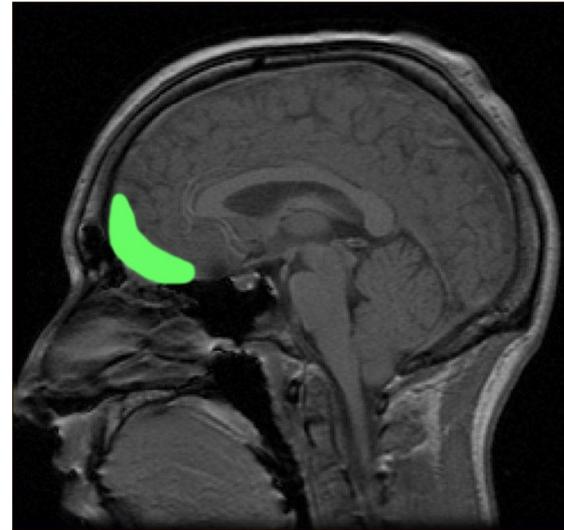
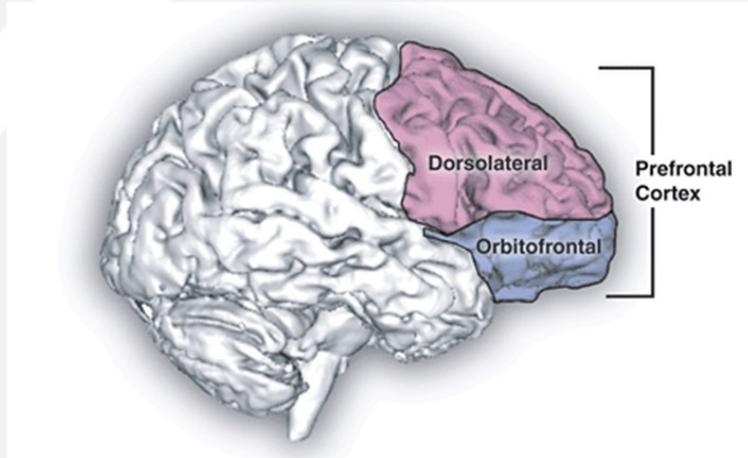
ADAM.

Prefrontal Cortex



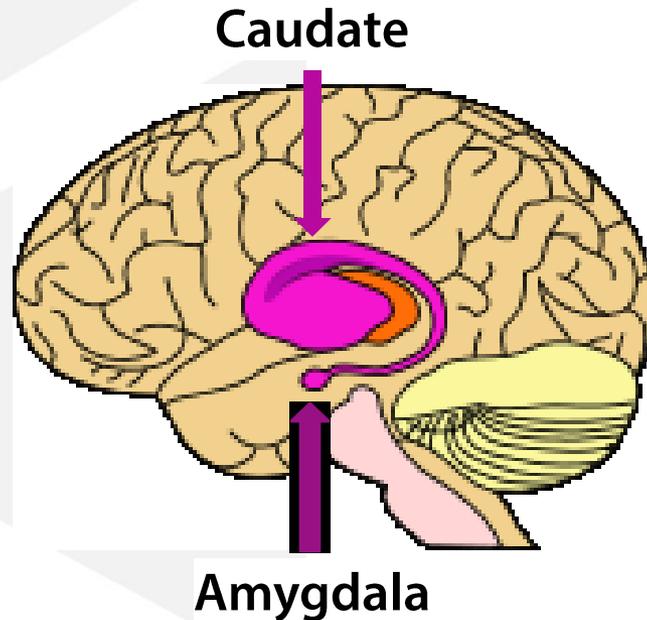
Prefrontal Cortex



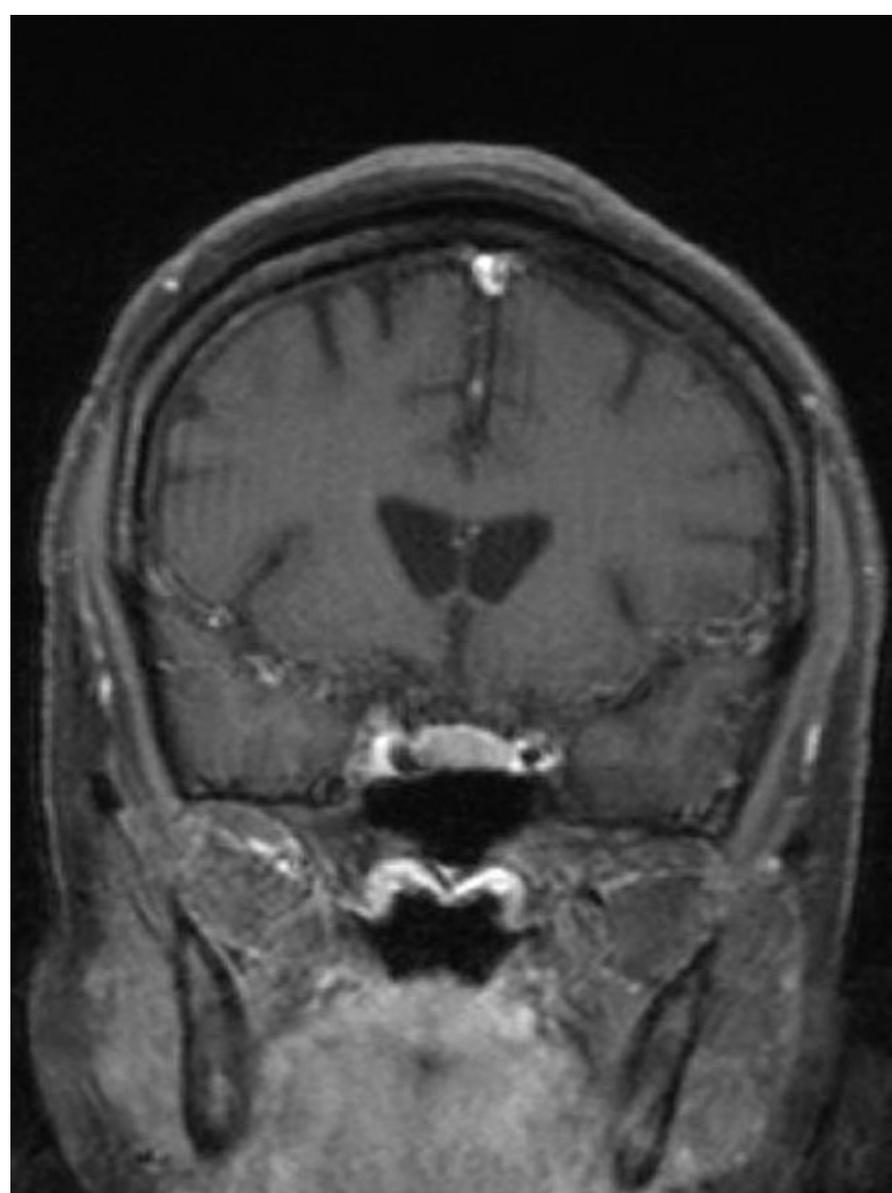
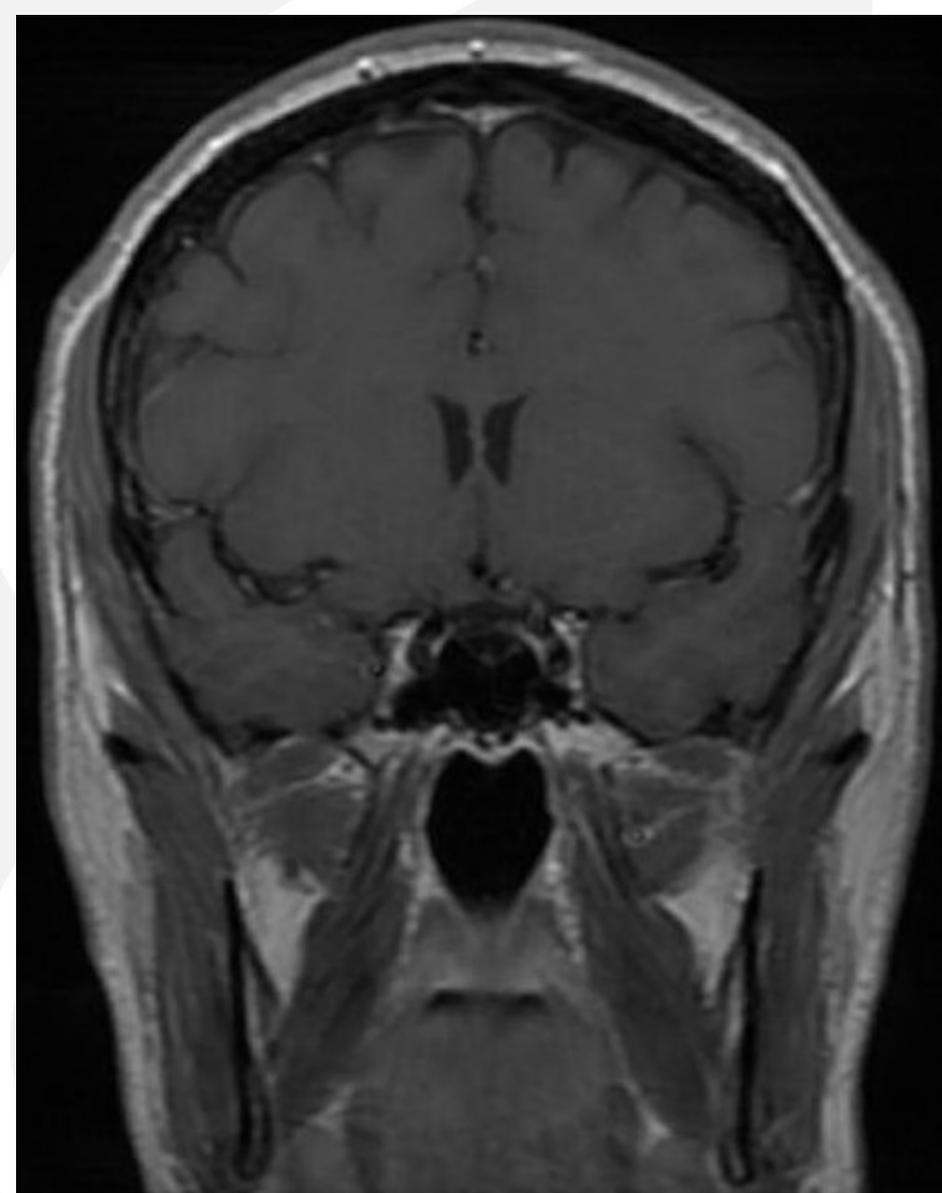


**OFC plays a key role in impulse control, monitoring ongoing behavior and socially appropriate behavior**

- **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

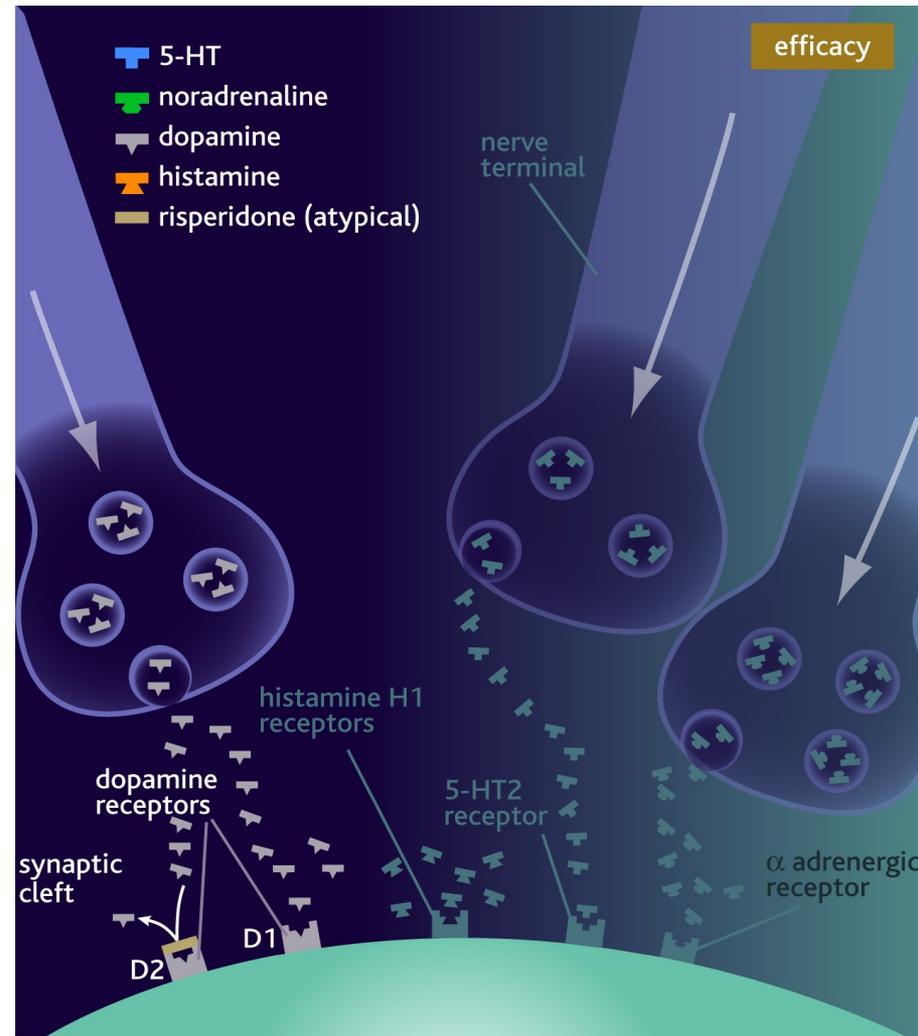
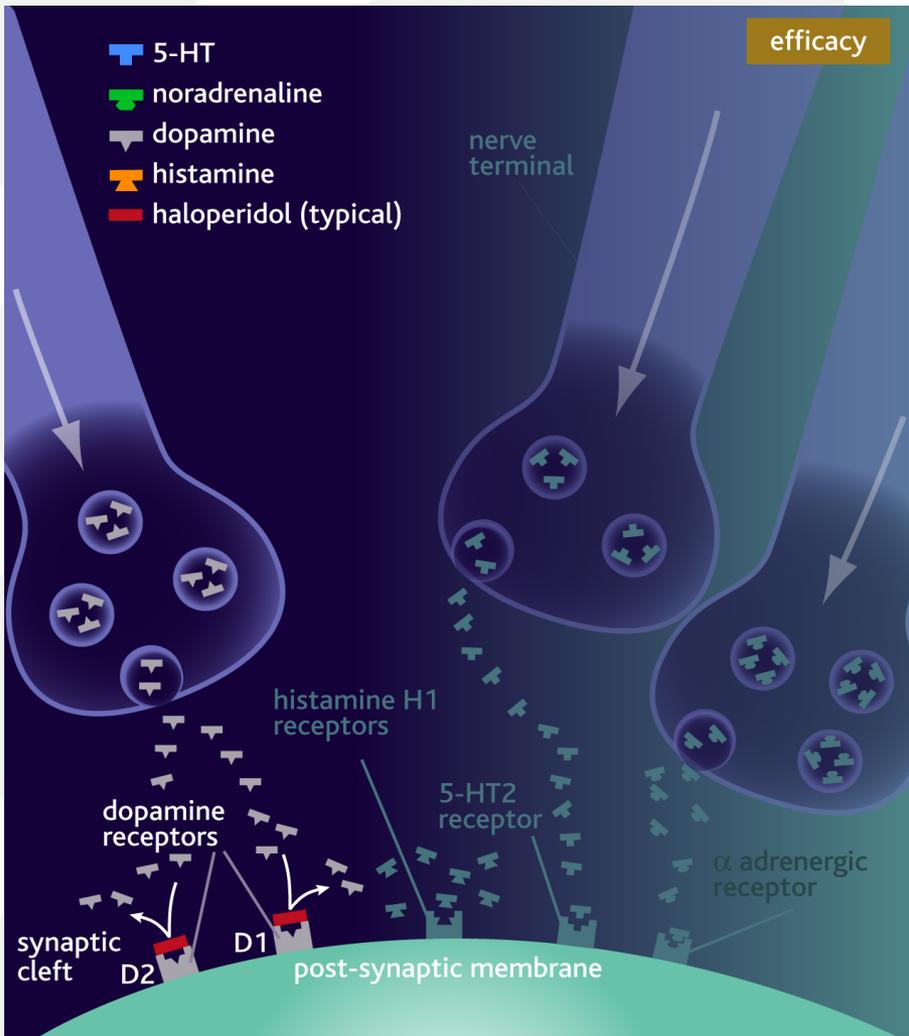
## MEDICATIONS:

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

# Aggression in Huntington's Disease

## MEDICATIONS:

### ❖ ANTI-PSYCHOTICS



© CNS Forum

# Aggression in Huntington's Disease

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

# Aggression in Huntington's Disease

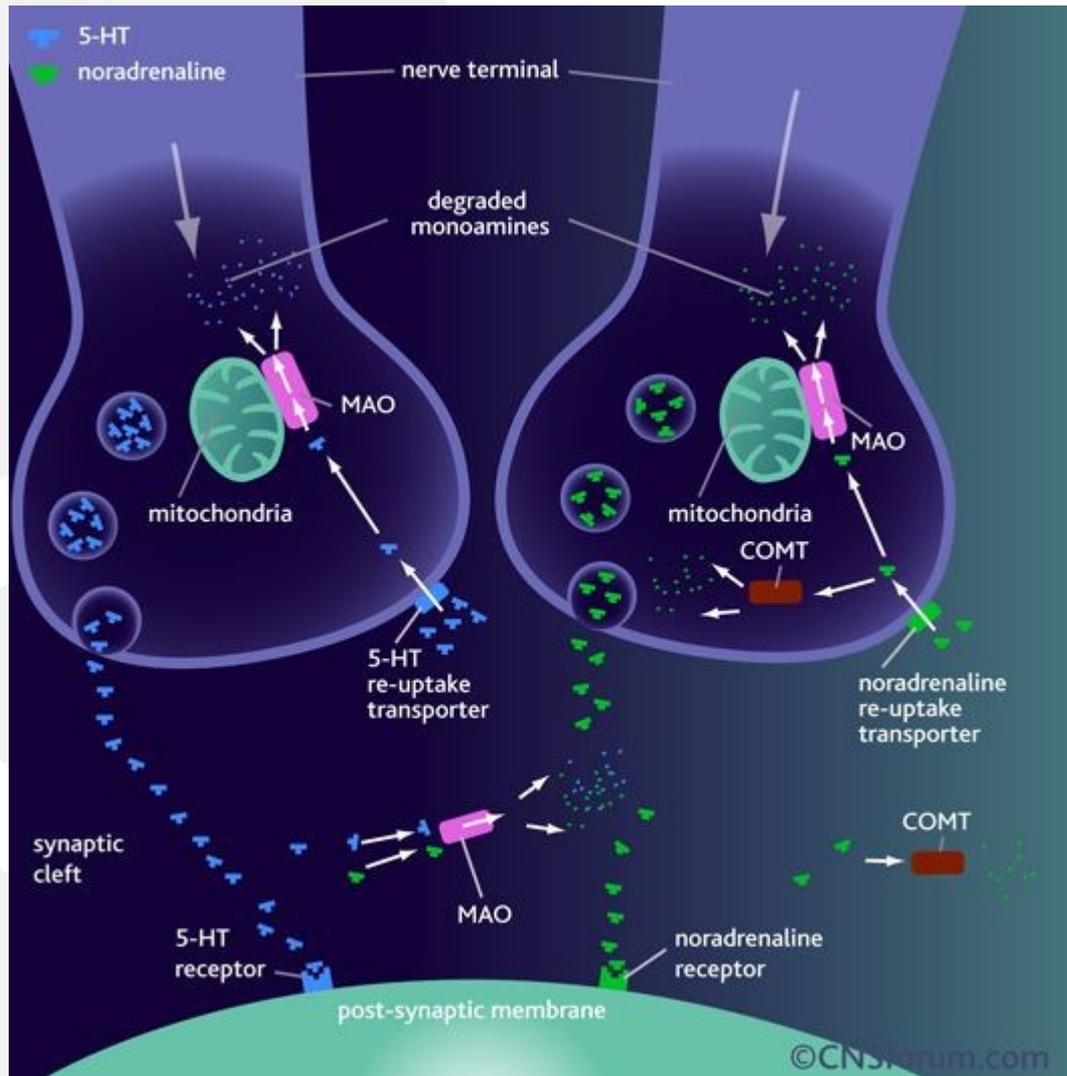
## MEDICATIONS:

### ❖ 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**



© CNS Forum

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

# Aggression in Huntington's Disease

## MEDICATIONS:

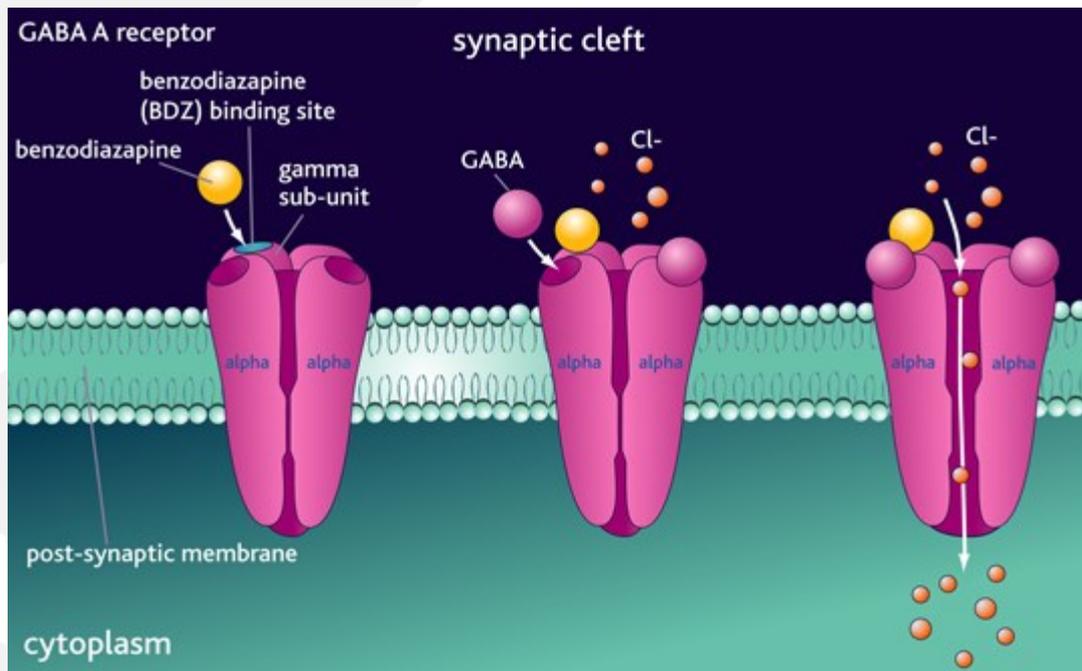
❖ ANTI-ANXIETY

# Aggression in Huntington's Disease

## ANTI-ANXIETY:

### ❖ BENZODIAZEPINES

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



© CNS Forum

# Aggression in Huntington's Disease

## BENZODIAZEPINES:

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

# Aggression in Huntington's Disease

## ANTI-ANXIETY:

### ❖ BUSPIRONE (BUSPAR)

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

# Aggression in Huntington's Disease

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

# 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/](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>

- Arik C. Johnson, Jane S. Paulsen, HDSA, 2014: Understanding Behavior In Huntington's Disease: A Guide For Professionals

- Martha A. Nance, Jane S. Paulsen, Adam Rosenblatt, Vicki Wheelock, HDSA 2011: A Physician's Guide to the Management of Huntington's Disease (3rd edition)

- Caroline A. Fisher, Katherine Sewell, Anahita Brown, Andrew Churchyard, Journal of Huntington's Disease, 3 (2014) 319-332: Aggression in Huntington's Disease: A systematic Review of Rates of Aggression and Treatment Methods

- Gill Livingston, Lynsey Kelly, Elanor Lewis Holmes, Gianluca Baio, et al, The British Journal of Psychiatry, 205 (2014) 436-442: Non-pharmacological interventions for agitation in dementia: systematic review of randomized control trials