Managing Aggression in Huntington’s Disease

Garima Arora, MD
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 *aggressi* – *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
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
"Hi."

"What's that supposed to mean!?!"
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
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-PYSCHOTICS:

<table>
<thead>
<tr>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Suppression of choreic movements</td>
<td>• Sedation</td>
</tr>
<tr>
<td>• May help with underlying depression/ apathy</td>
<td>• Metabolic Syndrome</td>
</tr>
</tbody>
</table>
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
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:

<table>
<thead>
<tr>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Treatment of underlying Depression or Anxiety or OCD</td>
<td>• GI Distress</td>
</tr>
<tr>
<td></td>
<td>• Headache</td>
</tr>
</tbody>
</table>
Aggression in Huntington’s Disease

MEDICATIONS:

- ANTI-ANXIETY
Aggression in Huntington’s Disease

ANTI-ANXIETY:

- **BENZODIAZEPINES**
  - Alprazolam (Xanax), Clonazepam (Klonopin), Diazepam (Valium), Lorazepam (Ativan)
# Aggression in Huntington’s Disease

## BENZODIAZEPINES:

<table>
<thead>
<tr>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Treatment of underlying Anxiety or OCD</td>
<td>• Disinhibition (may make agitation worse)</td>
</tr>
<tr>
<td>• Assistance with Sleep</td>
<td>• Confusion</td>
</tr>
</tbody>
</table>

- Treatment of underlying Anxiety or OCD
- Assistance with Sleep
- 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
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:**

<table>
<thead>
<tr>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>• May help with underlying depression/apathy</td>
<td>• Toxicity</td>
</tr>
<tr>
<td>• Divalproex helps with myoclonic hyperkinesia</td>
<td></td>
</tr>
<tr>
<td>• Lithium may help with motor function</td>
<td></td>
</tr>
<tr>
<td>• Lithium may reduce suicidality</td>
<td></td>
</tr>
</tbody>
</table>
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
- Arik C. Johnson, Jane S. Paulsen, HDSA, 2014: Understanding Behavior In Huntington’s Disease: A Guide For Professionals