

# Juvenile Onset Huntington's Disease

---

2008  
HDSA Annual Convention  
Jane S. Paulsen  
University of Iowa



# Juvenile HD

---

- ❑ HD age of onset 10% < 20; 25% > 50
  - ❑ Male transmission 70-80% JuvHD
  - ❑ CAG > 60 considered most common; 46% < 60 (42, 45, ...) > 80 repeats rare (< 5% JuvHD); largest CAG = 250
  - ❑ Avg range before diagnosis 9y (0-21)
  - ❑ 66% report psychiatric/behavioral 1st
  - ❑ 80% have psychiatric disturbance
-

# Medications

---

- ☐ Antidepressants
    - Tricyclics
    - SSRI
    - SNRI
  - ☐ Antipsychotics
  - ☐ Anticonvulsants
  - ☐ Mood stabilizers
  - ☐ Treatments for chorea
  - ☐ Treatments for spasticity/rigidity/dystonia
- 



# Non-medicine treatments

---

- ☐ Physical therapy
- ☐ Occupational therapy
- ☐ Speech therapy
- ☐ Counseling
- ☐ Behavior management
- ☐ Independent educational plan
- ☐ Alternative therapies



# Team Approach

---

- ☐ Social services
  - ☐ Financial planning
  - ☐ School programs
  - ☐ Physical supports
  - ☐ Medical symptom treatments
  - ☐ Nutrition/exercise schedules
  - ☐ Behavior Management Skills
  - ☐ Respite and Support for all Well-Being
- 



# Behavior Management Strategies

---



- ☐ ABC: Antecedent, Behavior, Consequences
- ☐ Operant/Classical Conditioning
- ☐ Parent Effectiveness Training
- ☐ Collaborative Problem-Solving
- ☐ Russell Barkley, Ross Greene



# Primary Principles:

## Behavior Management in Children

---

- ☐ Time perception
- ☐ Pick your battles
- ☐ Planning and Routine
- ☐ Intentional AND Unintentional reinforcement
- ☐ Follow-through



# Factors affecting Behaviors

---

## ☐ Fatigue

- Time of day
- Wellness
- Tiredness

## ☐ Frustration

- Task Complexity
- Organizational need
- Restraint needed for setting
- Communication or other limitations

## ☐ Competition

- Stimulation level in environment
- Evident or presumed alternatives

## ☐ Schedule of immediate/past consequences

**FABS**

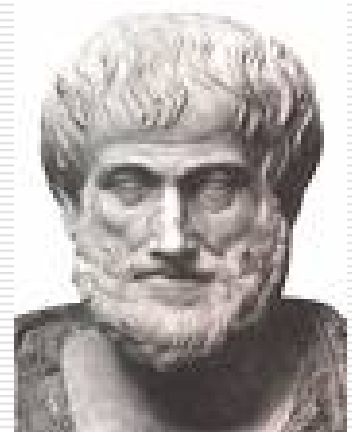


# Anger

---

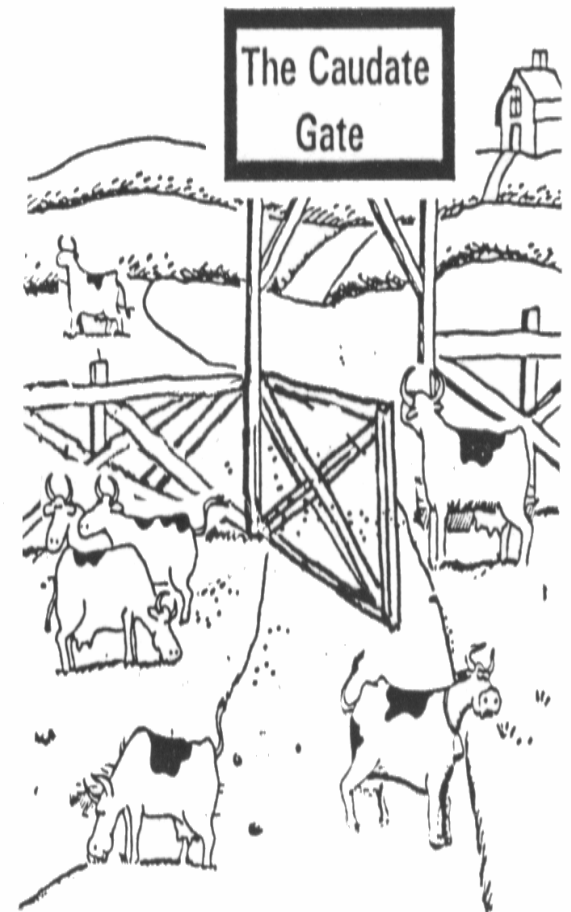
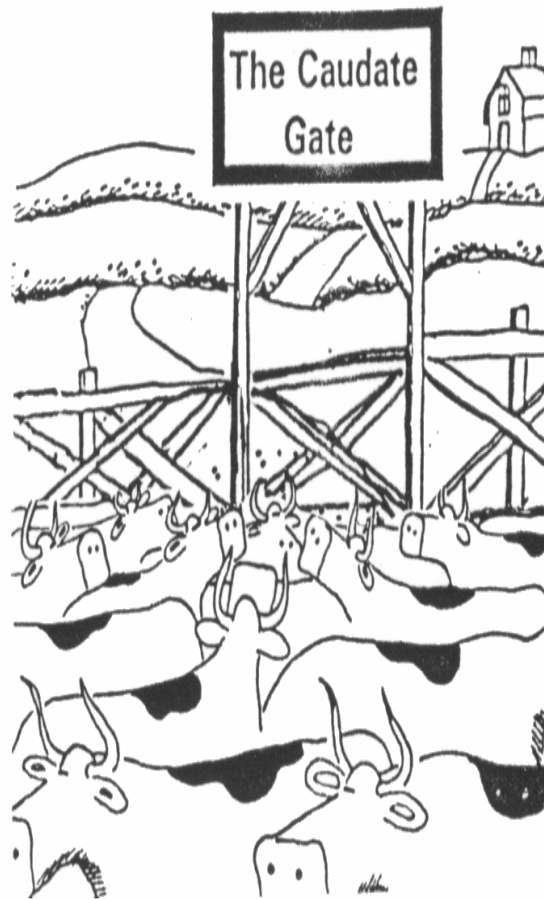
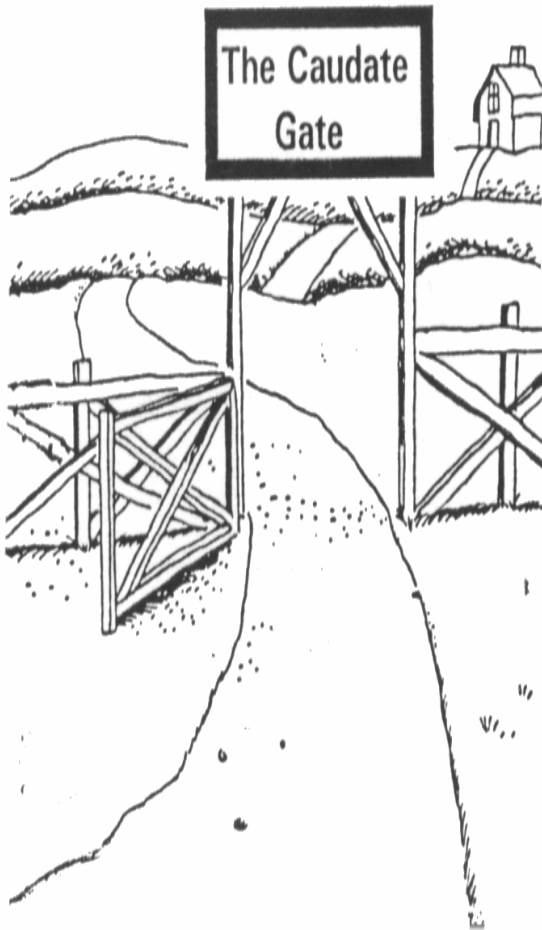
"Anyone can become angry,  
that is easy; but to be angry  
with the right person, to the  
right degree, at the right  
time, for the right purpose,  
and in the right way....  
.....this is not easy."

*Aristotle*



# "Gates" in the Brain

---



# What Happens When Gates Fall Out of Repair?

---

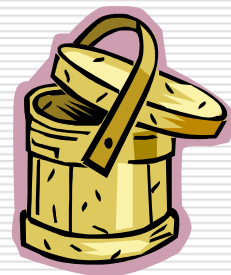
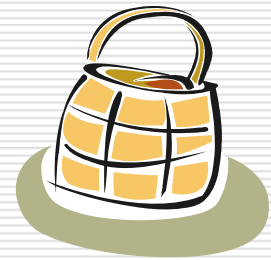
- ☐ Poor control of temper
  - ☐ Poor control of recall
  - ☐ Dysregulation of emotion and thinking
  - ☐ Confusion
- 



# Outbursts, temper,....

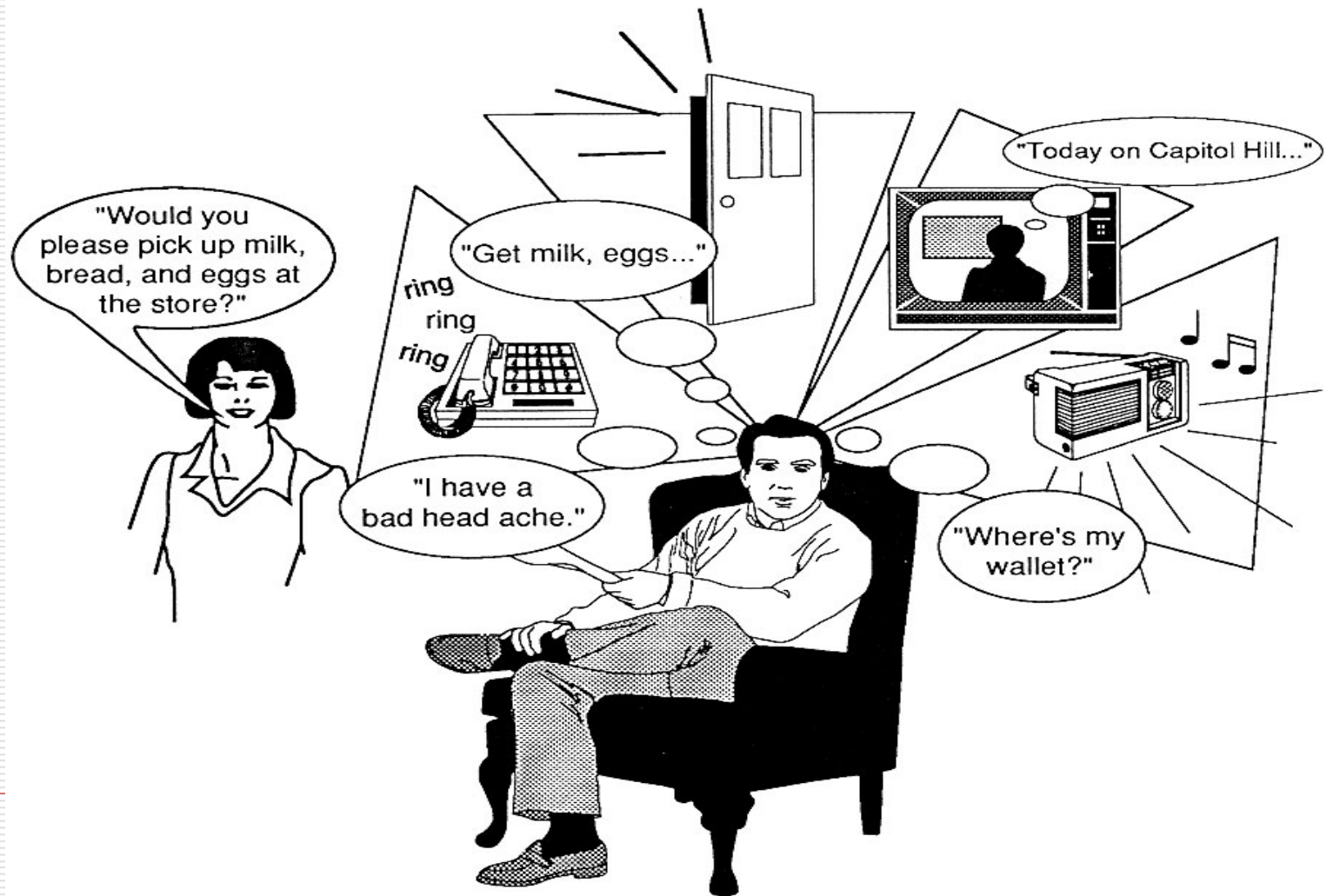
---

- ☐ Identify the antecedent or “trigger”
  - Bath example
- ☐ “Pick your fights”
  - Basket A – Safety
  - Basket B – Priority
  - Basket C – What does it matter?
- ☐ Acknowledge FEELINGS
- ☐ Assure safe environment(s)
- ☐ Check FABs



# Filtering, organizing, prioritizing

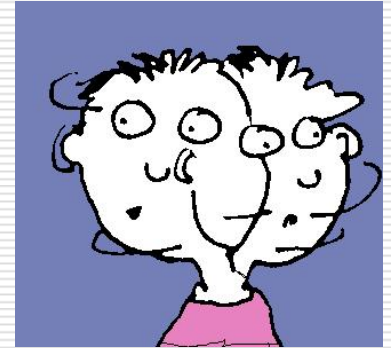
---



# Addressing Attention and “On-Task” Behavior

---

- ☐ Establish routine
- ☐ Baby steps
- ☐ Modify requests to parts that can be attained
- ☐ Celebrate accomplishments
- ☐ Reinforce Positively—go for it!



# Managing Obsessions



- ☐ Allow child to talk about interests
- ☐ Be creative integrating interests into school and home activities
- ☐ Gently shift to new topic or activity
- ☐ Recognize as symptom, not purposeful or willful
- ☐ Use prompts frequently to cue shift in activity or topic
- ☐ Agree to return to desired topic after "x"



# Maximizing Wellness and slowed Progression

---

Enriched environment

Low stress

Nutrition

Exercise

Wellness of caregiver

Wellness of family

---



# The rest of the family

---

- ☐ Take Breaks
  - ☐ Tag-team
  - ☐ Allow change and flexibility
  - ☐ Breathe
  - ☐ Forgive yourself and others
  - ☐ Each day is a new day
- 





