Juvenile Onset Huntington’s Disease

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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
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