Cognitive Rigidity and Atypicality:
When it’s more than ADHD

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Disclosures

- Dr. Coplan is author of Making Sense of Autistic Spectrum Disorders: Create the brightest future for your child with the best treatment options (Bantam-Dell, 2010), and receives royalties on its sale
- This presentation will include a discussion of off-label drug treatments
Case 1

CR is a four year old boy who presents to his primary care physician with parental concerns regarding “hyperactivity and tantrums.” The parents indicate that CR has always been a “high energy” child. He has been in pre-K for about 3 months; his teachers report “impulsivity and difficulty maintaining focus,” problems with task completion, and frequent temper tantrums.

Case 1

History: CR was conceived to a 29 y.o. G0 mother. Born at 35 weeks by vertex, vaginal delivery after PROM, BW 5 lb, discharged at 3 days of age. No postnatal issues. He is functioning at age level for language, self-care, gross- and fine-motor, and academic skills. His parents and a 2 year old sister are in good health. Family history is negative. Social history: Parents have been married for 6 years; both are employed.

Case 1

Examination

An alert, extremely active preschool child, with normal growth parameters, and PE. No dysmorphic features or focal neurological abnormalities. He speaks in full sentences, and seems to be of normal general intelligence.
Case 1

Medical Decision-Making
CR’s primary care physician makes a diagnosis of ADHD, based on the history of impulsivity and normal general intelligence and academic abilities, and prescribes amphetamine / dextroamphetamine tablets (Adderall®)

Case 1

Outcome
Within 48 hours, CR becomes extremely agitated, and experiences an uncontrollable “meltdown” because he cannot have access to an anticipated Christmas present, before Christmas arrives (in 2 weeks). He presents to the local ER, completely out of control, yelling, screaming, biting, and hitting.

Case 1

Examination (ER)
An alert, agitated, hyperventilating preschool child.
BP 100/70; P 100, HR 100
Skin is warm and most
Pupils moderately dilated, but reactive to light & accommodation
No focal findings on Neuro exam.
Patient insistently and repeatedly demands immediate access to his much-anticipated gift. Over the course of an hour, he gradually calms down.
Case 1

Medical Decision-Making
The patient is discharged from the ER; the parents are advised to see their PCP for referral to a mental health specialist (YOU).

What information would you like?

Case 1

Language: CR spoke single words at 12 months, 2-word phrases at 18 months, and full sentences by 24 months. He has a "broad vocabulary," and converses well with adults. He has a strong interest in septic systems and automobile transitions, and talks about these subjects incessantly.

Case 1

Behavior: CR can be very "strong-willed." He becomes angry if things do not work out the way he wants them to. He is very aware of routines, and becomes upset if they are changed. He has difficulty with transitions.
Case 1

Social: Over the past 24 months, his parents have become increasingly aware of CR’s difficulty socializing with other children. He will either disregard them, or attempt to dominate them, directing their play. Eye contact is good. He sometimes invades the personal space of others.

Case 1

Sensory-Motor: CR has food selectivity, limited to only about 10 things in his diet. He will refuse chicken nuggets if they have grill marks on them, for example. At the same time, however he eats nonfood objects such as facial tissues, and dog food. He has no fear, and will run away from his parents in the mall or on the playground.

Case 1

School report: “friendly and extremely intelligent... very high level of energy... difficult to get eye contact... children tend to ‘pull away’ from him... plays alone... he is very imaginative in his play, and will make his own ‘little world’... gets ‘one idea’ in his mind and will get obsessive about it for long periods of time.”
Case 1

Examination
An alert, active preschool child.
PE is normal
No focal neurological findings.
Subtle manneristic tensing of the body when excited.

Case 1

Cognitive-Behavioral Assessment
Mental Status:
• Eye contact: Good.
• Cooperation: Variable
• Attention span: Physical impulsivity, accompanied by verbal perseveration on automobile transmissions
  "The tracks got all apart... This could be a problem here!... We got a problem again. I'm going to get some help from the police... This is automatic... it's got 4-wheel-drive so I can go on the grass."

Case 1

• Vocabulary (PPVT-L): 5 yr 8 mo (SS 116).
  – Insists on naming all 4 pictures on each page ("stimulus driven").
  – Perseverates on transmissions:
    – "Is this tractor stick, or automatic?"
Case 1

- Gesell cube constructions and figure drawings: 3 ½ year level.
- Awkward crayon grip
- Difficulty imitating praxic finger gestures

Case 1

Difficulty with Theory of Mind and Central Coherence

Q: “Who is she?”
A: “An old lady….a grandmother.”
Q: “Who sent her that letter?”
A: “The postman”
Q: “No, I mean who wrote the letter?”
A: “I don’t know.”

Case 1

Diagnoses
1. Asperger Syndrome
2. ADHD
Case 1

Recommendations
- Language therapy (pragmatics)
- Social Skills
- Occupational Therapy
- Behavior Management
  - Cognitive rigidity
  - Anxiety
  - Disruptive Behavior
- Medication?

Case 1

Recommendations
- Language therapy (pragmatics)
- Social Skills

Case 1

- Behavior Management
  - Cognitive rigidity
  - Visual Schedules
  - Advance preparation for change
  - Anxiety
    - The Incredible 5-Point Scale (or similar)
    - Fidget toys, isometric exercise, mental imagery, CBT
  - Disruptive Behavior
    - Token Economy
Case 1

Medication?
- Cognitive Rigidity
- Dysregulation of Attention
- Dysregulation of Arousal

Neuropsychological Deficits in Persons with ASD

Abnormal Sensory Processing
Abnormal regulation of sleep
Abnormal regulation of attention
Abnormal regulation of arousal

Cognitive Rigidity
Abnormal regulation of arousal

Abnormal regulation of attention

Abnormal regulation of sleep

Abnormal Sensory Processing

Cognitive Rigidity

Agitation

Stereotypies

Routines

Atypical neuroleptics

D-2 agonists

SSRIs

Rigid

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Regulation of Attention

Attend to stimulus #1

Shift

Attend to stimulus #2

Abnormal Regulation of Attention - 1

- Perseveration
  - Inability to “Let go and shift”
  - Gets “stuck”
  - “Overattention Deficit Disorder”
- Compounds the effects of cognitive rigidity

Abnormal regulation of sleep

Abnormal sensory processing

Agitation, Aggression, SIB

Routines blocked

Abnormal regulation of arousal

Cognitive Rigidity

Abnormal regulation of attention

Rigid + Perseverative

SSRIs

SSRIs blocked

Abnormal Stereotypies

Abnormal Routines
Abnormal Regulation of Attention (Perseveration)

- Interventions
  - Verbal preparation for transitions
  - Visual Schedules
  - SSRIs
Abnormal Regulation of Attention - 2

- **Inattention**
  - Inability to focus
  - Impulsive
  - Distractible

**Interventions**

- Limited stimuli
- Short work periods
- Stimulants, alpha-2 agonists
  - Stimulants ➔ Cognitive Rigidity
  - Try guanfacine first, OR
  - Start an SSRI, then add stimulant
Inattention

- Beware of anxiety or perseveration masquerading as inattention
  - Perseveration on inner stimuli: “Inattentive”
  - Perfectionism: “Problems w. task completion”
  - Anxiety: “Rushes through work”

Abnormal regulation of attention

Abnormal regulation of arousal
  - Perseveration
  - Hypo-arousal
  - Hyper-arousal

Impulsive

Regulation of Arousal

Chronic hypoarousal

Fight or Flight Response

“Red Alert”
- Adrenaline
- Heart Rate
- Resp. Rate
- Combative

Calm & Relaxed

Red Alert
Abnormal regulation of attention
- Perseveration
- Inattention

Abnormal regulation of sleep
- Hypo-arousal
- Hyper-arousal

Routines
- Stereotypies
- Agitation
- Aggression
- SIB
- Sensory Seeking
- Sensory Overload

Impulsivity
- Hyperactivity

Atypical neuroleptics
- D-2 agonists

Disordered Sleep
- Melatonin

Sensory Dysfunction
- Sensory Seeking
- Sensory Overload

Disordered Sleep
- Melatonin

Rigid + Perseverative

Impulsive + Agitated / Disruptive
Abnormal regulation of arousal

Abnormal regulation of attention

(Perseveration)

(Inattention)

Cognitive Rigidity

Abnormal regulation of sleep

Abnormal Sensory Processing

Hyper-arousal

Hypo-arousal

Routines

Stereotypies

Agitation

Aggression

SIB

Sensory Overload

Impulsive

Agitated / Disruptive

Rigid + Perseverative

Disordered Sleep

Sensory Dysfunction

Sensory Seeking

SSRIs

Opiate antagonists

D-2 agonists

Impulsivity

Hyperactivity

Routines blocked

Abnormal regulation of attention

Atypical neuroleptics

D-2 agonists

Melatonin

SSRIs


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