MEDICATIONS IN X&Y CHROMOSOME VARIATIONS

MAKING MEDICATION DECISIONS & STRATEGIES TO SET AND MONITOR TREATMENT GOALS

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OUTLINE

OVERVIEW OF CONDITIONS TREATED WITH MEDICATIONS
MAKING DECISIONS ABOUT MEDICATIONS
SYMPTOMS TREATED WITH MEDICATIONS
NUTS AND BOLTS:
   STARTING MEDICATIONS
   DEVELOPING TREATMENT GOALS
   MONITORING RESPONSE AND SIDE EFFECTS
INTRODUCTION TO MEDICATION CLASSES
RESOURCES
CASE EXAMPLES
I’m not endorsing any specific medication, brand, or company

I have received research funding for Fragile X research from:
Seaside, Roche, Neuren, Alcobra

I have consulted for:
Novartis, Zynerba, Ovid

All medications should be discussed with your doctor

We cannot make specific recommendations without evaluating a patient
XYY, XXX, XXYY, XXY, etc

Differences in Brain Development / Function

**INCREASED RISK FOR:**

**Cognitive / Learning Differences**
- Developmental Delay
- Learning Disability (LD)
- Intellectual Disability (ID)

**Social Development Differences**
- Social immaturity
- Autism Spectrum Disorder (ASD)

**Behavioral / Emotional Differences**
- Short Attention Span
- Distractibility
- Hyperactivity
- Impulsivity
- Anxiety
- Depression

**Sensory Sensitivities**
- Irritability / Aggression
- Obsessive-Compulsive Behaviors
- Repetitive / Stereotypic Behaviors

**ADHD**
TREATMENT PLAN

- Academic supports
- OT
- Speech
- Behavior Therapy
- Medications
- Other
WHEN SHOULD MEDICATIONS BE CONSIDERED?

- When behavioral/emotional difficulty is significantly impacting:
  - Health and Safety
  - Academics / Learning
  - Therapies
  - Participation in the family / community
  - Social development / Social Interactions

When child is expressing / demonstrating distress about the emotion/behavior
COMMON PARENTAL CONCERNS

???
COMMON PARENTAL CONCERNS

• I don’t want my child to become dependent on them or addicted to them
• I don’t want my child to be “drugged”
• I don’t want to change his/her personality
• I am worried medications will change the way the brain develops
COMMON PARENTAL CONCERNS

• I’m worried about side effects
• He/she will have to take it for life
• Family member/friend took them and had a bad reaction
• I read about a person who took them and...
• I want my child to learn to manage his/her own behaviors without needing medications
WHEN SHOULD MEDICATIONS BE CONSIDERED?

When behavioral/emotional difficulty is significantly impacting:

▪ Health and Safety
▪ Academics / Learning
▪ Therapies
▪ Functioning in the family / community
▪ Social development / Social Interactions

When child is expressing / demonstrating distress about the emotion/behavior
WHEN SHOULD MEDICATIONS BE CONSIDERED?

When behavioral/emotional difficulty is significantly impacting:

- **Health and Safety**

  Injuries: To self and others (Aggression / Self-Injurious Behaviors)

  Risk of Injuries: To self and others (Aggression / Self-injury / Running off / Impulsive Behaviors)

  Health:
  - Poor growth/Weight gain (attention span w/meals, restricted eating due to sensory or anxiety concerns with food / eating environment)
  - Constipation / GI issues (anxiety / refusal to poop at school)
WHEN SHOULD MEDICATIONS BE CONSIDERED?

When behavioral/emotional difficulty is significantly impacting:

- Academics / Learning
- Therapies

- How much time is spent in managing “behavior” (attention span, anxiety, etc.) versus engaged in the learning/school environment or therapy?

- Are they making the rate of progress that they are capable of in academic / therapy environments?

Is their academic placement or availability of therapy limited by their behavior?
WHEN SHOULD MEDICATIONS BE CONSIDERED?

When behavioral/emotional difficulty is significantly impacting:

- Functioning in the family / community
- Causing significant conflict with siblings / family
- Not able to participate in family / community events
- Family not participating in family / community events
WHEN SHOULD MEDICATIONS BE CONSIDERED?

- When behavioral/emotional difficulty is significantly impacting:

  - **Social development / Social Interactions**
    
    Impulsivity / Attention Span affecting social interactions / play
    
    - “Annoying” or “Not fun to play with”
    
    - “Long term memory” of negative behaviors in kids
      
      “She’s the girl who….”
      
      “He’s the kid who…”

  - Anxiety / Social Withdrawal / Selective Mutism
    
    - Limited ability to have / engage in social interactions
    
    - Limited ability to have / engage in social activities
WHEN SHOULD MEDICATIONS BE CONSIDERED?

▪ When behavioral/emotional difficulty is significantly impacting:

▪ **When child is expressing / demonstrating distress about the behavior**
  ▪ “Why is this happening?”
  ▪ “I don’t like…” / “I can’t help it”
  ▪ Self-injury
  ▪ Suicidal ideation / attempt
CONSIDERATIONS FOR STARTING TREATMENTS

- BENEFITS
- RISKS / COSTS
### BENEFITS

- Behavior might improve
- Overall functioning may improve at home, in school / work, in community
- Improvement in social interactions / with others / social opportunities
- Feeling that you are doing everything you can to help your child

### RISKS

- Side Effects / Long term effects
- Medication may not work
- Medication may worsen other behavior
- Feeling that it is the “easy way out”, you are not helping your child to learn to regulate behavior on their own

### COSTS / Appointments

- Lab draws / Monitoring distressful

### TAKING MEDICATION

- Avoid side effects and cost
- Avoid worries of side effects
- You may find other treatments / supports to help the problem
- If behaviors continue to be a problem, you can consider them later

### NOT TAKING MEDICATION

- Behavior can continue to cause problems / stress for patient, in family, in school / work setting
- Behavior may get worse
- Behavior may limit opportunities
- Behavior may limit progress in school / therapies
- Behavior may cause health / safety problems
TIMING OF STARTING MEDICATIONS

Symptoms present

Symptoms causing mild-to-moderate impairment/problems

Symptoms causing moderate-to-severe impairment/problems

CRISIS

Academic Failure
Severe safety concerns / injuries
Kicked out of school
Mental Health Hospitalization
TIMING OF STARTING MEDICATIONS

Medications as “the last option”

- Safety concerns
- Impact of prolonged severe behavioral difficulties and stress on:
  - Self (Child / Adult with XY variation)
  - Family relationships
  - Social relationships
  - Academic placements / relationships
  - Long term physical and mental health
CONSIDER MEDICAL PROBLEMS CONTRIBUTING TO BEHAVIOR DIFFICULTY

Consider / Rule-Out:

Pain – ear infections / dental problems
Constipation / GI symptoms
Headaches
Vision / Hearing problems
Seizures
Thyroid problems
Sleep apnea
DECISION: MEDICATION TRIAL

Now what?
IDENTIFYING TARGET BEHAVIORS

What is the goal of medication treatment?
What would you like to see improve?
What behavioral / emotional difficulty is leading to impairment in overall functioning?
Differences in Brain Development / Function

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INCREASED RISK FOR: ADHD
IDENTIFYING TARGET BEHAVIORS

7 year old boy with XXY (Albert):

“Behavior is getting worse, the school keeps calling me because of his behavior, going to the grocery store or restaurant with him is horrible and we don’t go anymore, he almost got hit by a car yesterday in the parking lot, he got kicked out of speech therapy”
IDENTIFYING TARGET BEHAVIORS

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CONSIDERATIONS FOR MEDICATIONS:

SAFETY???
ACADEMICS / THERAPY???
FAMILY / COMMUNITY FUNCTIONING???

WHICH MEDICATION?

WE NEED MORE DETAILS / SPECIFICS
IDENTIFYING TARGET BEHAVIORS

7 year old boy with XXY (Albert):

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CONSIDERATIONS FOR MEDICATIONS:

SAFETY: YES
ACADEMICS / THERAPY: YES
FAMILY / COMMUNITY FUNCTIONING: YES

WHICH MEDICATION?

WE NEED MORE DETAILS / SPECIFICS
TARGET BEHAVIORS

“The school keeps calling me because of his behavior”

Why?

1. He can’t sit through group activities / “circle time”, the teacher always needs to redirect him to his work, he gets out of his seat, he grabs belongings of other students

OR

2. He has meltdowns every time he transitions and “gets stuck”, he’s overwhelmed in the classroom, when he doesn’t want to do something he walks out of the room, becomes disruptive with peers
TARGET BEHAVIORS

“Going to the grocery store or restaurant is horrible”

Why?

1. He can’t sit still / hyperactivity / touches or grabs everything, runs down the aisle, yells when he can’t get something he wants

OR

2. Gets overwhelmed in busy places, doesn’t like new places / change in routine, asks over and over when we are going home, then melts down after 5-10 minutes
TARGET BEHAVIORS

“He almost got hit by a car in the parking lot”

Why?

1. He ran off when he saw the ice cream store he wanted go in

OR

2. He ran off when he saw he was at the dentist office
TARGET BEHAVIORS

“He got kicked out of speech therapy”

Why?

1. He likes her but he can’t pay attention long enough to do what the therapist wants him to do, he keeps getting out of his chair, touching all her materials, she spends all of the visit trying to get him to get through a couple exercises or redirecting him

OR

2. He becomes agitated in that small room, she talks loudly and it’s painted in bright colors, he’s frustrated and overwhelmed within 5-10 minutes, and then he melts down or “we lose him” for the rest of the session
TARGET BEHAVIORS

TARGET BEHAVIOR: Attention Span / Hyperactivity / Impulsivity

MEDICATION CONSIDERATION: ADHD Medication

MONITOR RESPONSE TO MEDICATION:

- Improved attention span at school and during therapy (from 1-2 minutes currently to ___ minutes)
- Ability to tolerate going to the grocery store / restaurant (i.e. less fidgety / able to tolerate short trips without a meltdown)
- Decreased impulsivity (less grabby, less interrupting others, less running off/unsafe impulsive behaviors)
TARGET BEHAVIORS

Boy #2

TARGET BEHAVIOR: Anxiety
MEDICATION CONSIDERATION: Anxiety Medication
MONITOR RESPONSE TO MEDICATION:

Better able to tolerate transitions
Decreased vocalizations / rocking when overwhelmed
More flexible with changes, new places and changes in routine / fewer meltdowns
Fewer “panic” reactions leading to safety risks
MEDICATION TREATMENT PLAN

Identify target behaviors and determine how to measure them

▪ Frequency
▪ Duration
▪ Intensity
▪ Formal/Standardized vs. Observational/”Informal”
MEDICATION TREATMENT PLAN

Treatment Plan:

▪ TEAM
  ▪ TEAM CAPTAIN – Individual taking the medication
  ▪ TEAM Members – 1-4 people to help monitor response to medication
    ▪ Parent/caretaker, teacher, friend, therapist / psychologist, siblings
MONITORING RESPONSE TO TREATMENT

**Monitoring Tools:**
- Standardized Questionnaires
  - ADHD symptoms
  - Others…

**Completed by:**
- Parents
- Teachers
- Therapists / Other providers
**MONITORING RESPONSE TO TREATMENT**

<table>
<thead>
<tr>
<th>BEHAVIOR</th>
<th>never</th>
<th>occasionally</th>
<th>often</th>
<th>very often</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Does not pay attention to details or makes careless mistakes;</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>for example, homework.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Has difficulty attending to what needs to be done.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>3. Does not seem to listen when spoken to directly.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>4. Does not follow through when given directions and fails to finish</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>things.</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>5. Has difficulty organizing tasks and activities.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>6. Avoids, dislikes, or does not want to start tasks that require</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>ongoing mental effort.</td>
<td></td>
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<tr>
<td>7. Loses things needed for tasks or activities (assignments, pencils,</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>or books).</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>8. Is easily distracted by noises or other things.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>9. Is forgetful in daily activities.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>10. Fidgets with hands or feet or squirms in seat.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>11. Leaves seat when he/she is supposed to stay in his/her seat.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>12. Runs about or climbs too much when he/she is supposed to stay</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>seated.</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>13. Has difficulty playing or starting quiet games.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>14. Is “on the go” or often acts as if “driven by a motor”.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>15. Talks too much.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>16. Blurs out answers before questions have been completed.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>17. Has difficulty waiting for his/her turn.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>18. Interrupts or bothers others when they are talking or playing</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>games.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>19. Argues with adults.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>20. Loses temper.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>21. Actively disobey or refuses to follow an adult’s requests or rules.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>22. Bothers people on purpose.</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Directions: Each rating should be considered in the context of what is appropriate for the age of your child and should reflect that child’s behavior in the last 6 months. Is this evaluation based on a time when the child □ was on medication □ was not on medication □ not sure?
Monitoring Tools:

2. Behavior Logs

- Formal behavior logs in the school setting
- Parent-developed logs / tracking systems
  - At time of each incident
  - Daily ratings each night
    (Green, Yellow, Red days)
- Communication notebooks with schools
- OT / Speech / PT Therapy reports
FINDING A DOCTOR

Primary Care Doctor
Referral from Primary Care Doctor
  Developmental Pediatrician
  Child Adolescent Psychiatry
  Developmental Neurology
Referrals from local Family Groups / Autism groups
AXYS Clinic & Research Consortium Clinic
FINDING A DOCTOR

Who can’t prescribe medications?*
Psychologists
Teachers
Therapists (OT, Speech, PT)

*But, they can all be very helpful as part of team monitoring effects of medication
DIFFERENT STYLES OF DOCTORS

Authoritative
“Father / Mother Figure”
Dictator

Collaborative
Partner
Consultant
Team Member

ALSO:
DIFFERENT STYLES OF PATIENTS
HELP YOUR DOCTOR BE A GOOD DOCTOR

Come to appointment with:

- Ideas of specific treatment goals
- Specific examples of difficult behaviors
- Behavior logs (if available)
- Written questions (2-4)
- Written input from teachers
- Copies of reports from any medical evaluation / psychology or therapy progress results, IEPs since last visit
STARTING MEDICATIONS

Start low, go slow

One change at a time
- No other changes in meds, therapies or environment
- Summer / school breaks

Start

2-3 weeks, reassess

4-6 weeks
IMPORTANT QUESTIONS TO ASK

What are the common side effects?

What are the rare side effects to watch for?

If side effects happen, what should we do?

How/who do I contact if I am concerned about the effects of the medications?
  • During the day?
  • After hours / weekends?

Does the medication need to be taken every day?

What should I do if he/she misses a dose?

What if I run out or want to stop – is it dangerous to stop abruptly?
MONITORING MEDICATIONS

Follow-up Visits
Necessary at certain frequencies
  Range from every 1-6 months to yearly
Allow doctor to see and examine patient
  Physical Examination (other medical problems / findings)
  Observe response to treatment
  Side effect monitoring
  Lab monitoring
Can be done without patient present (pediatrics)

Phone calls
OK for adjustments / updates/ mild difficulties
Sometimes necessary if physically far from provider
Not billable
Behavioral / Emotional Differences

Short Attention Span
Hyperactivity
Impulsivity

ADHD Medications
Stimulants
Nonstimulants

Anxiety

SSRI Medications
Alpha agonists

Irritability / Aggression
Self-injurious Behavior

SSRI Medications
Atypical Antipsychotics
Mood Stabilizers

Obsessive-Compulsive Behaviors (OCD)

SSRI Medications
ATTENTION PROBLEMS ARE NOT ALWAYS ADHD…

Also consider:
- Learning / Cognitive Impairments
- Anxiety
- Sensory sensitivities

Medical considerations:
- Vision
- Hearing
- Thyroid problems
- Seizure disorders
- Sleep apnea
MEDICATIONS FOR ADHD

Stimulants
- Methylphenidate
  - Ritalin, Concerta, Metadate, Focalin
  - Dexedrine / Adderall / Vyvanse

Nonstimulants
- atomoxetine (Strattera)
- guanfacine (Intuniv)
- clonidine (Catapres, Kapvay)
- bupropion (Wellbutrin)
"I couldn't help it. My lack of serotonin made me do it."
SSRIS

- Anxiety
- Shyness
- Withdrawal
- Worries / Fears
- Panic Attacks
- Irritability
- Depression

Mechanism: Increase brain serotonin levels

Examples:
citalopram (Celexa)
escitalopram (Lexapro)
sertraline (Zoloft)
fluoxetine (Prozac)
SSRIS – POTENTIAL BENEFITS

less irritability, improved mood, fewer outbursts with anxiety trigger
able to transition more easily
less self-picking, nailbiting, other behaviors
Fewer worries, perseverations
able to tolerate changes / frustrations more easily
able to be more comfortable in social settings
some show increased speech if selective mutism or anxiety component to low speech
less fixations and compulsive behaviors
Less tearfulness / emotional sensitivity “easy to cry”
SSRIS - SIDE EFFECTS

activation/hyperactivity/disinhibition - wild/odd behaviors – about 10-15%
appetite changes
insomnia - usually wears off
nausea - not common
decreased sexual drive/impotence - rarely an issue
OTHER MEDICATION TREATMENTS

Atypical Neuroleptics (Atypical Antipsychotics)
- Risperdal (risperidone)
- Abilify (aripiprazole)
- Seroquel (quetiapine)
- Others

Mood Stabilizers
- Lithium
- Depakote
- Trileptal
- Lamictal
QUESTIONS....