

# MEDICATIONS IN FRAGILE X: PART A

## INTRODUCTION TO MEDICATIONS & STRATEGIES TO SET AND MONITOR TREATMENT GOALS

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Children's Hospital Colorado

# OUTLINE

**CONFERENCE MEDICATION SERIES**

**MAKING DECISIONS ABOUT MEDICATIONS**

**BEHAVIORS TREATED WITH MEDICATION IN FXS**

**NUTS AND BOLTS:**

**STARTING MEDICATIONS**

**DEVELOPING TREATMENT GOALS**

**MONITORING RESPONSE AND SIDE EFFECTS**

**INTRODUCTION TO MEDICATION CLASSES**

**RESOURCES**

**CASE EXAMPLES**

# DISCLAIMER / DISCLOSURES

**I'm not endorsing any specific medication, brand, or company**

**I have received research funding from:**

**Seaside, Roche, Neuren, (Alcobra)**

**I have consulted for:**

**Novartis**

**All medications should be discussed with your doctor**

**We cannot make specific recommendations without evaluating a patient**

**I will not write any prescriptions today for children or their parents**

# MEDICATION TALK SERIES

**7/17 Thurs 9:45-11:00**

**Part A: Introduction and Strategies to set and monitor treatment goals**

Dr. Nicole Tartaglia

**7/17 Thurs 4-5:15**

**Part B: Hyperactivity, Inattention, Sleep Problems, and Aggression**

Dr. Ave Lachiewicz and Dr. Carol Delahunty

**7/19 Fri 11:15a-12:30p**

**Part C: Medications for Anxiety, Irritable Behaviors, Aggression**

Dr. Liz Berry-Kravis

**Others:**

**7/17 Thurs 8:30-9:30am: Targeted Treatments: Drs. Berry Kravis and Hagerman**

**7/18 Fri 11:15am-12:30pm: Use of Medications to Treat Hyperarousal: Dr. Lachiewicz**

**7/18 Fri 11:15am-12:30pm: Data from the Seaside Trial (Arbaclofen): Dr. Berry Kravis**

**7/20 Sun 9:00am-10:15am: Medication Panel – Come and Get Answers to your Medication Questions:**

Drs. Berry-Kravis, Hagerman, Tartaglia, Erickson, Lachiewicz, Feldman

**Many other posters and abstracts**

**Fragile X Syndrome**

**Differences in Brain  
Development / Function**

**Cognitive / Learning Differences**

**Developmental Delay  
Developmental Disability  
Intellectual Disability (ID)  
Learning Disability (LD)**

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

**Behavioral / Emotional Differences**

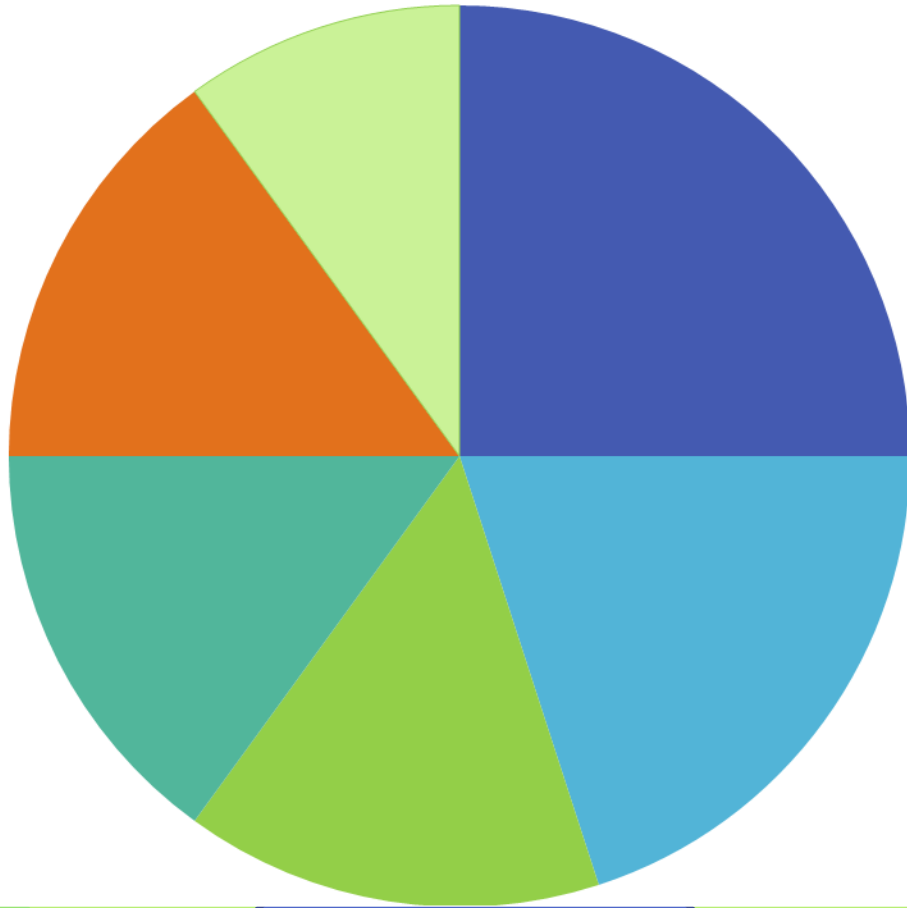
**Short Attention Span  
Hyperactivity  
Impulsivity** ———— **ADHD**

**Anxiety  
Hyperarousal  
Sensory Sensitivities  
Irritability / Aggression  
Self-injurious Behavior  
Obsessive-Compulsive Behaviors  
(OCD)  
Repetitive / Stereotypic Behaviors**

# MEDICATIONS AS PART OF A TREATMENT PLAN



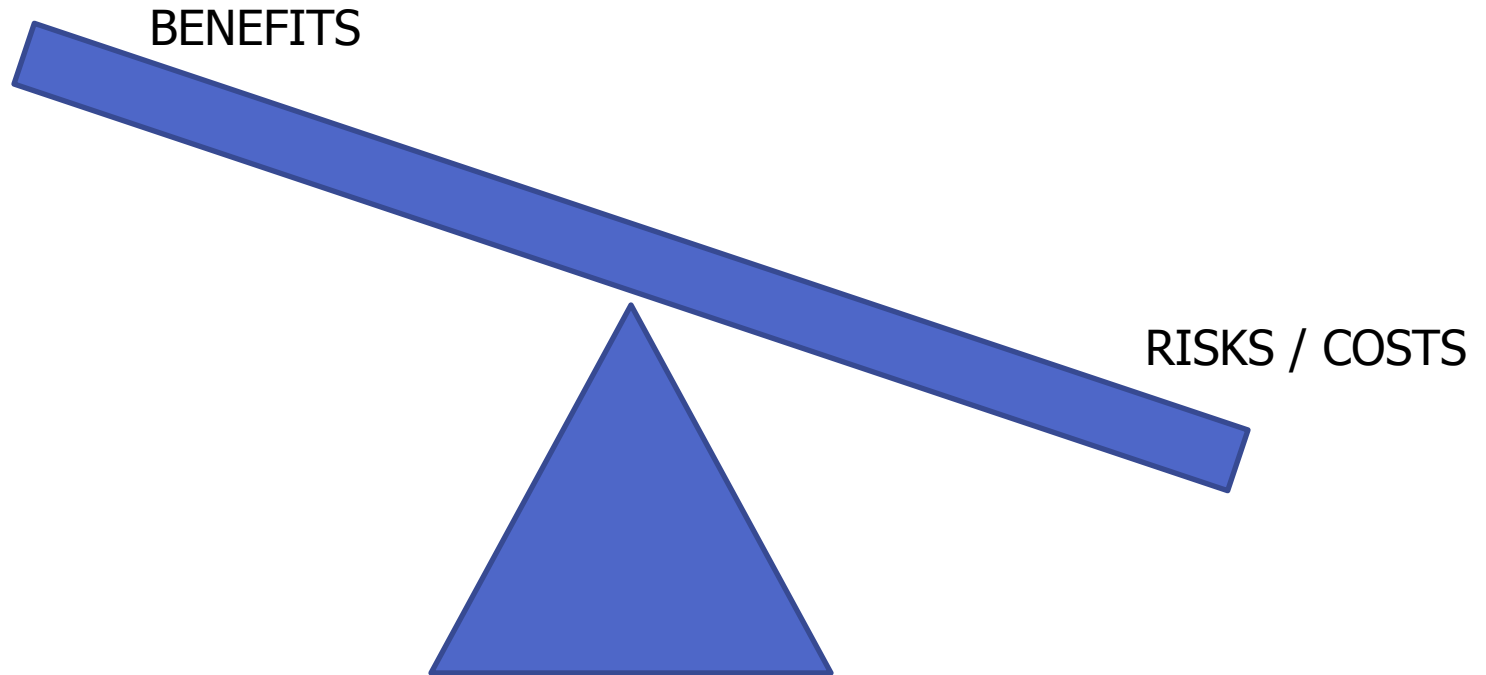
- Academic supports
- OT
- Speech
- Behavior Therapy
- Medications
- Other



- Academic supports
- OT
- Speech
- Behavior Therapy
- Medications
- Other


**Family / Home Environment**

# CONSIDERATIONS FOR STARTING TREATMENTS






# 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...
- 

# BENEFITS

# RISKS

TAKING MEDICATION

**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**

**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**

NOT 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**

**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**

# BENEFITS

# RISKS

TAKING MEDICATION

A large, empty rectangular box with a thin green border, intended for listing the benefits of taking medication.A large, empty rectangular box with a thin green border, intended for listing the risks of taking medication.

NOT TAKING  
MEDICATION

A large, empty rectangular box with a thin green border, intended for listing the benefits of not taking medication.A large, empty rectangular box with a thin green border, intended for listing the risks of not taking medication.

# WHEN SHOULD MEDICATIONS BE CONSIDERED?

- When behavioral/emotional difficulty is significantly impacting:
  - Health and Safety
  - Academics / Learning
  - Therapies
  - Social development / Social Interactions
  - Participation in the family / community
- When child is expressing / demonstrating distress about the 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 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:
  - 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:
  - Participation 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:
- **When child is expressing / demonstrating distress about the behavior**
  - “Why is this happening?”
  - “I don’t like...” / “I can’t help it”

# TIMING OF STARTING MEDICATIONS

**Symptoms present**



**Symptoms causing mild-to-moderate impairment/problems**



**Symptoms causing moderate-to-severe impairment/problems**



**CRISIS**

**Severe safety concerns / injuries**

**Kicked out of school**

# TIMING OF STARTING MEDICATIONS

## **Medications as “the last option”**

- Safety concerns
- Impact of prolonged severe behavioral difficulties and stress on:
  - Self (Child / Adult with FXS)
  - Family relationships
  - Social relationships
  - Academic placements / relationships

# 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**

# 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?**



**Fragile X Syndrome**

**Differences in Brain  
Development**

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(OCD)  
Repetitive / Stereotypic Behaviors**

# IDENTIFYING TARGET BEHAVIORS

**7 year old boy with FXS (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

**7 year old boy with FXS (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”**

**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 rocks and makes noises that are disruptive**



# TARGET BEHAVIORS

**“Going to the grocery store or restaurant is horrible”**

**Why?**

**1. He can't sit still / hyperactivity / touches or grabs everything, then melts down when redirected or when he can't have something he wants**

**OR**

**2. Extreme fear of shopping carts, gets overwhelmed in busy places, doesn't like new places / change in routine, then melts down after 5-10 minutes**



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# 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 running around the room, touching all her materials, she spends all of the visit trying to get him to get through one exercise 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



## Boy #1

**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 / Hyperarousal**

**MEDICATION CONSIDERATION: Anxiety (SSRI)? Abilify? Clonidine?**

**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"



# TARGET BEHAVIORS / SYMPTOMS VS. TARGETED TREATMENTS

**Target Behavior / Symptom:** The behavior or symptom being targeted for improvement with a specific treatment (medication or therapy)

## **Examples:**

- Attention span
- Aggression
- Anxiety

**Targeted Treatment Medication:** Medication being used to target the underlying neurobiological abnormalities in FXS

## **Examples:**

### **Clinical Trial (Research Study) medications**

mGluR5 Antagonists / GABA agonists:

Arbaclofen, mavoglurant (Novartis), Roche

- **FDA Approved Off-label use:** Minocycline, Acamprosate

# 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

Medication Treatment Plan

¶

TEAM CAPTAIN: → → → → TEAM MEMBERS: → → → → ¶

→ → → → → → → → → → → ¶

→ → → → → → → → → → → ¶

→ → → → → → → → → → → ¶

¶

Target Behavior / Symptom: ¶

Medication Name / Dose: ¶

¶

How will I know it is working? (Specific Target behaviors / symptoms) ¶

1. ¶

2. ¶

3. ¶

Side Effects to Watch for: ¶

1. ¶

2. ¶

3. ¶

If I have concerns call: ¶

→ 911 – Emergency ¶

→ Clinic Phone: ¶

→ After Hours / Doctor on Call: ¶

For Refills call: ¶

# MONITORING RESPONSE TO TREATMENT

## Monitoring Tools:

**Standardized Questionnaires**

**ADHD symptoms**

**ABC (Aberrant Behavior Checklist)**

**Others...**

**Completed by:**

**Parents**

**Teachers**

**Therapists / Other providers**

# MONITORING RESPONSE TO TREATMENT

Parent's Name: \_\_\_\_\_ Today's Date: \_\_\_\_\_ Child's Name: \_\_\_\_\_ Age: \_\_\_\_\_

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?

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

# MONITORING RESPONSE TO TREATMENT

## 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

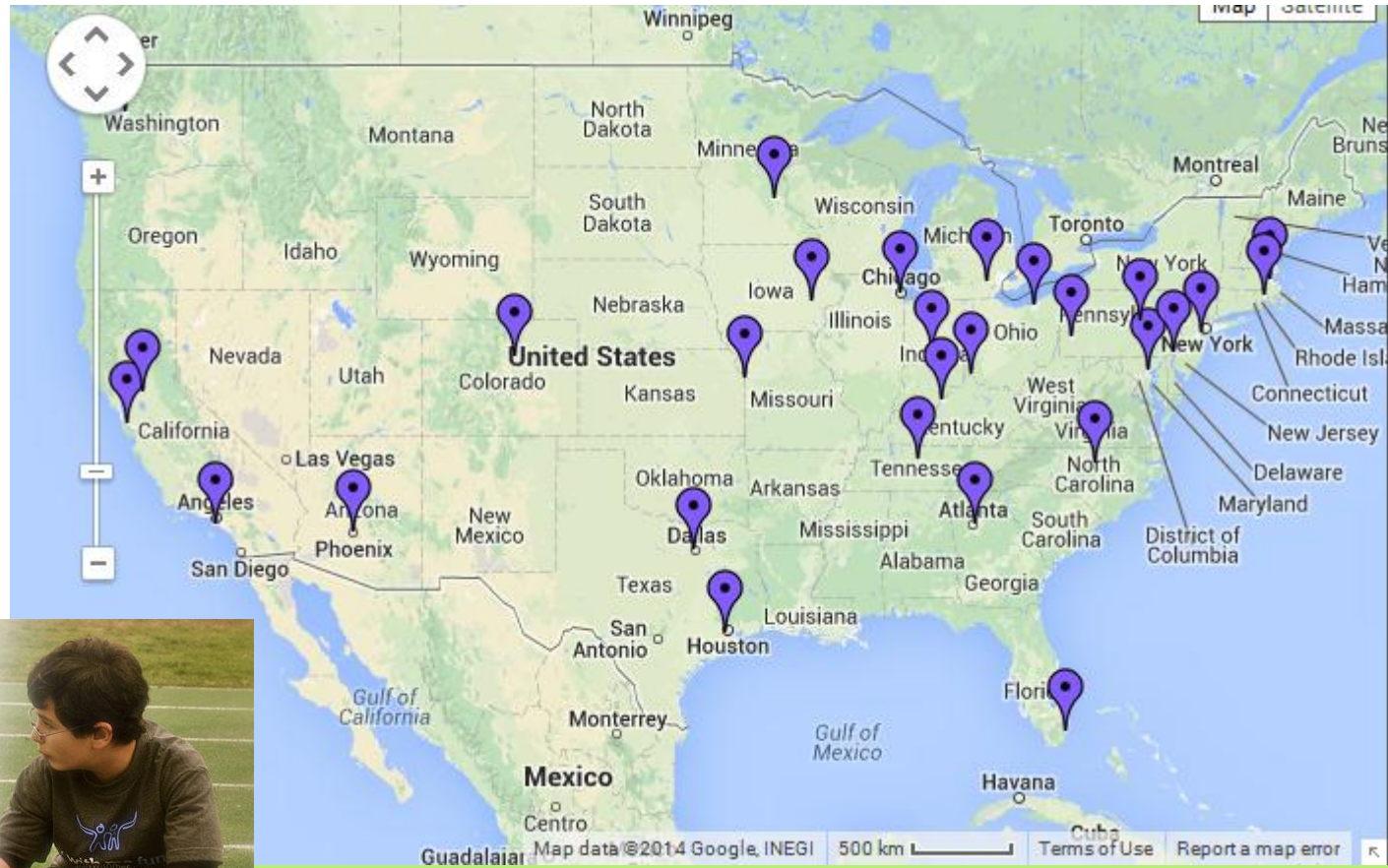
(Green, Yellow, Red days)

Communication notebooks with schools

OT / Speech / PT Therapy reports

# FINDING A DOCTOR

## Fragile X Clinics:



# FINDING A DOCTOR

## Who can prescribe medications?

- Primary Care Provider –
  - Pediatrician, Family Practice Doctor, PA/NP
- Child – Adolescent Psychiatrist
- Developmental-Behavioral Pediatrician
- Child Neurologist

## 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**

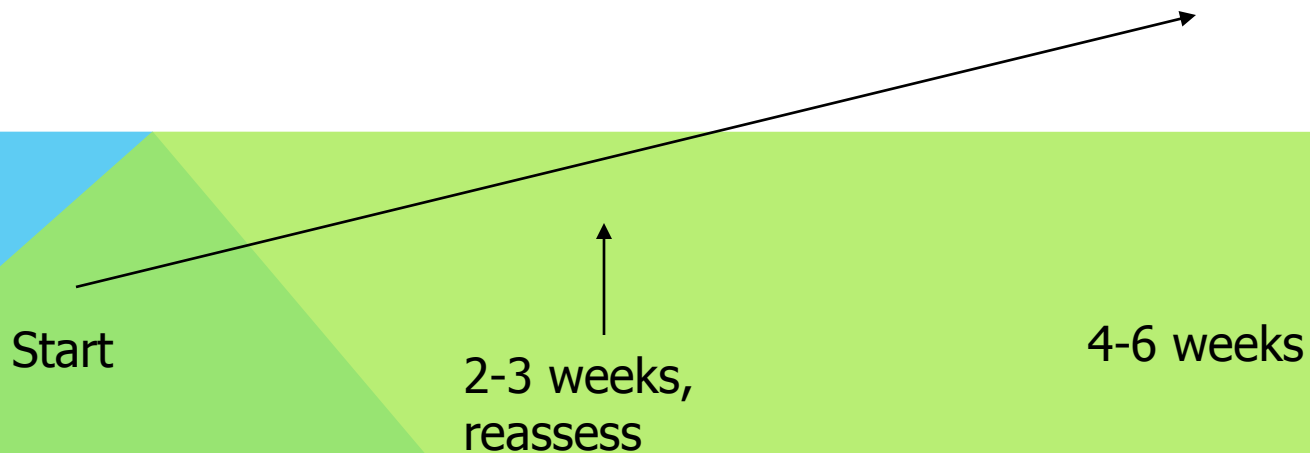
**Copies of reports from any medical evaluation / psychology or therapy progress results, IEPs since last visit**

# STARTING MEDICATIONS

**Start low, go slow**

**One change at a time**

- No other changes in meds, therapies or environment
- Summer / school breaks



# 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

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(OCD)  
Repetitive / Stereotypic Behaviors**

## Behavioral / Emotional Differences

Short Attention Span  
Hyperactivity  
Impulsivity



ADHD Medications  
Stimulants  
Nonstimulants

Anxiety



SSRI Medications  
Alpha agonists

Hyperarousal  
Sensory Sensitivities



SSRI Medications  
Alpha agonists

Irritability / Aggression



Self-injurious Behavior

SSRI Medications  
Atypical Antipsychotics  
Mood Stabilizers

Obsessive-Compulsive Behaviors (OCD)



SSRI Medications

Repetitive / Stereotypic Behaviors



SSRI Medications  
Treatment needed?

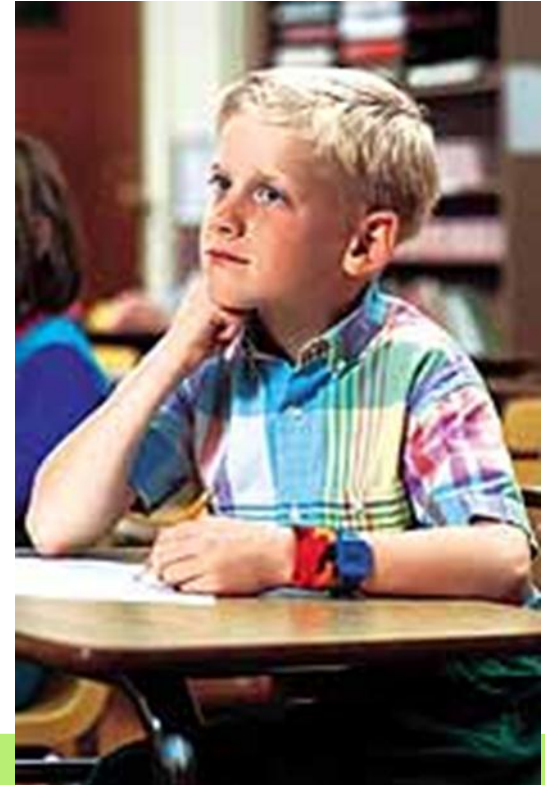
# MEDICATIONS FOR ADHD

## Stimulants

- Methylphenidate
  - Ritalin, Concerta, Metadate, Focalin
  - Short acting
  - Long acting
- Dexedrine / Adderall / Vyvanse

## Nonstimulants

- Atomoxetine (Strattera)
- Guanfacine (Intuniv)
- Clonidine (Catapres, Kapvay)





*Kellogg's*

# RITALIN-O's

Fortified with Methylphenidate HCL

I Eat Them  
With Bananas  
heehhehehe!!

KIDS GO  
**CRAZY**



For  
**Ritalin-O's!**

NET WT. 18OZ (1LB 2OZ)



# ATTENTION PROBLEMS ARE NOT ALWAYS ADHD...

Also consider:

- Learning / Cognitive Impairments
- Anxiety
- Sensory sensitivities

Medical considerations:

- Vision
- Hearing
- Thyroid problems
- Seizure disorders
- Sleep apnea

# MEDICATION TREATMENTS

## **SSRI medications**

- Celexa (citalopram)
- Zoloft (sertraline)
- Prozac (fluoxetine)
- Lexapro (escitalopram)

## **Atypical Neuroleptics (Atypical Antipsychotics)**

- Risperdal (risperidone)
- Abilify (aripiprazole)
- Seroquel (quetiapine)
- Others

## **Mood Stabilizers**

- Lithium
- Depakote
- Trileptal
- Lamictal

# RESOURCES



**National Fragile X Foundation:**

<http://www.fragilex.org/treatment-intervention/medication>

**FXCRC Consensus Documents:**

<http://www.fragilex.org/treatment-intervention/consensus-on-clinical-practices/>

**Medication Guidelines:**

[http://www.fragilex.org/wp-content/uploads/2012/08/Medications\\_for\\_Individuals\\_with\\_Fragile\\_X\\_Syndrome2012-Oct.pdf](http://www.fragilex.org/wp-content/uploads/2012/08/Medications_for_Individuals_with_Fragile_X_Syndrome2012-Oct.pdf)

**Autism Speaks Medication Decision Guide:**

<http://www.autismspeaks.org/science/resources-programs/autism-treatment-network/tools-you-can-use/medication-guide>

Consensus of the Fragile X Clinical & Research Consortium on Clinical Practices

Medications for Individuals with Fragile X Syndrome



Autism:  
Should My Child Take  
Medicine for Challenging Behavior?



A Decision Aid for Parents  
of Children with Autism  
Spectrum Disorder



# QUESTIONS....

