

International Rett Syndrome Foundation Conference 2009

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[Please note that “child” can also refer to an adult with Rett syndrome]

Sometimes the behavior is just the expression of the child's internal mood or medial response. It might be that a chemical imbalance in the brain from Rett Syndrome or seizure activity (or both). Behavioral techniques and medications are listed below by areas of behavioral difficulty.

When behavioral techniques don't work well enough and the child is suffering impaired, then the family can consider using a medicine.

I recommend that a family consider a medicine if behavioral treatment isn't working and a child has one of the three criteria for using a medicine and no medical cause could be identified.

1. The child is suffering
2. The child is a danger to herself or others
3. The child is impaired in school or at home

All medication trials are “trials ”. It is up to the parents to decide if they see enough benefit that they think is worth the risk, hassle, or cost of using a medicine.

When I try to determine what medicine to use , I ask questions about sleep, behavior, energy, appetite, moodiness, irritability, crying spells, concentration, self-injury (including if skin injury, eye problems result), aggression.

Medical information:

I also ask what medical and psychiatric illnesses run in the family and what medical illnesses the child has – including allergies, sinus infections, ear infections, (acid) reflux, bone fracture and seizure disorder. If a behavior is new and sudden, it is more likely to be due to a medical cause.

Sleep

Sleep problems can be addressed behaviorally sometimes but the sleep problems in Rett syndrome can be very difficult to treat.

Sleep problems can be from seizure activity.

Clonidine can be helpful for sleeping. Medicines and the family of "neuroleptics" have a side effect of sleepiness but also appear to help correct the sleep pattern. These neuroleptics aren't often used for a sleep problem alone but may be a good choice if self-injury or aggression is present. Medicines in this class include Haldol (haloperidol), Risperdal (risperidone), and Seroquel (quetiapine), Geodon (ziprasidone), Abilify (aripiprazole), Orap (pimozide), Zyprexa and Zyprexa Zydis (olanzapine).

These medicines are used to treat bipolar disorder, schizophrenia, and confused states when ill. They all can cause temporary muscle stiffness (EPS) and a movement disorder that might not go away ever (Tardive Dyskinesia). They can also cause neuroleptic malignant syndrome.

You can obtain “blood levels” that measure how much is in the body for Risperdal Risperdal (risperidone) and Zyprexa / Zyprexa Zydis (olanzapine).

Treatment for self injury and aggression.

Ear infections, allergies, and sinus infections can lead to self-injury.

Head hitting can damage the ear drum and eyes.

The neuroleptic family helps self injury and aggression.

Behavioral techniques can be very helpful.

Obsessions and compulsions (Obsessive compulsive disorder).

Individuals with Rett syndrome frequently have behaviors or movements they repeat over and over. These include hand wringing, opening and closing doors and drawers, turning light switches on and off, repeating other behaviors.

Medicines that can be tried for repetitive behaviors.

Medicines in the family of Prozac (fluoxetine), Luvox (fluvoxamine), Celexa (citalopram), Zoloft (sertraline), Escitalopram (Lexapro).

Anxiety Symptoms.

Anxiety is often seen in Rett syndrome.

The antidepressants and benzodiazepines - Valium (diazepam), Ativan (lorazepam) can help anxiety and maybe selective mutism. Lorazepam is sometimes used for calming or sedation when medical procedures are performed.

Autism spectrum disorders, pervasive developmental disorders (I'll call these all "autism" here).

Autism is an artificial name that was created to describe a series of symptoms seen together: social difficulties, communication difficulties, and unusual or repetitive behaviors. It is not a separate disorder.

In 1 out of every 10 people we can find the cause of the autism. Some of these causes are fragile X syndrome and Rett syndrome.

Knowing the symptoms of autism can be helpful to Rett families, doctors, other health specialists, and the schools system, so that the best treatment and school programs are put in place. Most school districts now have a much better understanding of autism and are using the educational systems that have been put in place that help address the sensitivities and needs of individuals with autism. Individuals with Rett syndrome may benefit from being in classrooms that were created for individuals with autism. These classrooms usually are smaller in size and emphasize visual instructions. For example, the school may use a picture board that shows the activities of the day. This helps the

child to transition from one activity to another without becoming upset. An autism school program also usually has social skills training. This is when the teachers or speech therapists work to increase the individual's ability to interact with others. These autism programs also have a better understanding of behavioral techniques that work for children with Rett syndrome.

Self Injury

Arm splints (sleeves) can be used to decrease self-injury and hand-wringing. When used for hand-wringing, the splint can often be placed on only one arm. When a splint is used, often the child will pay more attention to her surroundings.

Skin Protection:

Biting and mouthing of hands and shoulders frequently occur. Arm splints may help. Neoprene (wet suit) material can be bought and shown to make a hand or shoulder cover. Vaseline is non-toxic and adds a barrier to the saliva.

Special situations that cause a person with Rett syndrome to become upset

Many individuals with Rett syndrome become easily agitated by sounds and confusion. Head phones with a favorite song playing may be helpful.

Mood disorders: Depressive disorders (depression) and Bipolar disorder (manic depressive disorder).

The word "depression" is a misleading name for the illness. Many people have depressive disorders but never actually feel sad. They may instead be irritable, cranky, or fussy. Children can have depression and complain of feeling bored and angry but never feel sad.

The medicines for depression are antidepressants like Prozac or Wellbutrin that help to put brain chemistry that might be abnormal because of Rett syndrome. St. John's Wort might be used but may have the same side effects as other antidepressants.

Some people feel depressed in the winter (have a seasonal depression) and cell light boxes on can be helpful or antidepressants can be used. Exercise can also be helpful for depression.

If there is a family history of manic depression, also known as bipolar disorder, antidepressants and light boxes can make the bipolar active. St. John's Wort can also function as an antidepressant and may make bipolar disorder active.

Bipolar disorder is an illness in which people can feel angry irritable and at other times overly happy. Many people with RETT SYNDROME look like they have bipolar disorder. This is even if they don't have a family history of bipolar disorder.

Bipolar disorder may respond to lithium, Depakote (divalproex sodium), Depakene (valproic acid), Trileptal (oxcarbazepine), Tegretol (carbamazepine), and Lamictal

(lamotrigene). For all of these medicines, you can obtain “blood levels” that measure how much is in the body. They all have some potential side effect. The makers of Trileptal say that you do not need to get blood tests to look for problems of the liver and blood cells like you do with the other medicines.

Attention deficit hyperactivity disorder ("hyperactivity", ADHD)

ADHD can present with both attention problems and hyperactivity or with just attention problems. Girls tend to have attention problems without as much high activity. The attention problems include having a hard time hearing when they are told to stop doing bad behaviors.

Medicines that can help are methylphenidate (Ritalin, Metadate, Concerta), dextroamphetamine (Dexedrine, Adderall [mixture of Dexedrine salts], and clonidine. Methylphenidate (Ritalin, Metadate, Concerta), dextroamphetamine (Dexedrine, Adderall [mixture of Dexedrine salts] cause a decrease in appetite. Strattera is a new medicine that lasts through the day and evening but may cause liver

Blood tests: Emla cream can be prescribed to numb the skin of the arms before blood tests. You can use plastic wrap (like Seran wrap) to help keep it on. Duct tape may help as well if it is placed toward the ends of the plastic wrap.

Applied Behavior Analysis

- **Strategies to assess and treat problem behaviors**
- **Effective in producing behavior increasing communication and decreasing self-injury and aggression.**

Key elements

- Evaluation of situation
- Documentation of progress
- Use of well-researched techniques

What is a *behavior problem*?

- Maladaptive, observable behavior that persists or increases, impairs functioning
- Caused by inadvertent learning
- Triggered by events that bring out the behavior (antecedents)
- Maintained by reinforcement things that increase the chance of the behavior happening in the future

Examples of behavior problems

- aggression
 - hitting, kicking, biting, scratching, head butting, pinching, slapping, choking, hair pulling
- self-injury
 - head banging, head hitting, self-biting, hair pulling, eye poking, self-scratching, skin picking
- property destruction
 - ripping, breaking

- disruptive behavior
 - throwing objects, screaming, yelling, banging on surfaces
- pica
 - ingesting inedible items
- elopement
 - running from designated/supervised area
- incontinence
- noncompliance
 - passive refusal
- dangerous behaviors
 - climbing, standing on furniture, playing with electrical cords, fire setting, seeking knives/sharp objects

Choosing a behavior to change

- Is the behavior *dangerous*?
 - What is the *frequency* of the problem behavior (How often does it happen)?
- *How long has the behavior been present?*
- Will behavior change increase future *skill development*?
 - Does behavior lead to *family stress*?
 - Some behaviors are more difficult to change **than others**.
 - *Cost* of behavior change (Time and money)?

What is Next?

- Define the behavior
 - **Aggression: hitting, kicking**
 - **Self-injury: arm biting, hand biting**

Measuring the behavior

- Goal of Applied Behavioral Analysis (ABA)
 - measure (*behaviors in actual situation*)
 - *evaluate change; don't continue ineffective program; don't discontinue effective program prematurely*

Types of Data Collection

- **Frequency**
 - record each occurrence of the behavior
 - Ex. Aggression per hour, self-injury per day
 - » can compare each day to another even if data collected at different times
 - **Sample Behavior Data Collection Sheet**
- Name:
- Please record the frequency of the targeted behaviors listed below.

■ Time	SIB	AGG	DIS	YELLING	
■ 7:00 - 7:30		0		YES	NO
■ 7:30 - 8:00	0	0	0	YES	NO
■ 8:00 - 8:30	0	0	0	YES	NO
■ 8:30 - 9:00	0	0	0	YES	NO
■ 9:00 - 9:30				YES	NO

Functional Assessment

- Develops *hypothesis* of function of problem behavior (trying to see how the child “use” the behaviors? What causes it to keep happening?)

Functional assessment questions

- When does the behavior always occur?
- When does the behavior never occur?
- What happens before the behavior?
- What happens after the behavior?
- What is the individual trying to communicate to you?
- Does the individual seem to want a reaction from you following the behavior?
- Does the individual use the behavior to get out of doing things?
- Does the individual have more of the behavior when he/she needs to wait for something?
- Does the individual engage in the behavior even if no one else is around?
- Does the individual engage in the behavior in a repetitive fashion?

Why is Function Important?

- Understand why it’s happening, so you don’t make accidental mistakes
 - Example 1. Being upset with the child may make the behavior worse when the child just wants your reaction.
 - Example 2. Time-out may make the behavior worse if the child just wants to get out of doing something

Ways to respond to behaviors that a child does to get attention

- Ignore (*look away, make no comments, walk away*)
- Block and ignore (*prevent behavior from occurring but make no comment or eye contact*)
- Time-out (*remove child to isolated area but provide no comment, eye contact, or other attention*)
- **Teach child to request attention**
- **Functional Communication Training**
- **Picture Exchange Communication Systems**
- **Offer praise if there are no problem behaviors**
- **Offer attention to specific positive behaviors**

If a behavior happens to try to get something

- State “no” once and continue with activity
- Provide no comment or other attention
- Teach child to request specific item or “more”
- Use a picture schedule to show when an item is available (if possible)
- Use item as a reinforcer for other behaviors

If a behavior happens to avoid an activity:

- Tell the child to continue the behavior
 - provide no comment or other attention
- Stop if necessary, but return to task once calm
- Teach child to request break or help, if possible (such as with pictures)
- Allow child to earn breaks
- Allow child to earn other powerful reinforcers

Behavior continue because of a physical sensation

- Includes self-stimulation, relief from discomfort
- Block and ignore behavior
- Provide alternative sources of stimulation
- Address physical problems
- Teach toy play skills if absent

Treatment Goals

- **Increase appropriate behavior**
 - **Reinforce (when there are no bad behaviors)**
 - **Teach and reinforce appropriate “replacement behavior”**
 - Make more available the things or interactions that improve behavior
- **Decrease problem behavior**
 - **Brief, time-limited consequences**

Added when initial interventions insufficient