



# **Overview and treatment of extreme behavioral problems in SLOS**

**Elaine Tierney MD  
Kennedy Krieger Institute  
Johns Hopkins University School of Medicine  
Baltimore, MD**

**Autism spectrum disorder (ASD):  
autism, pervasive developmental  
disorder, Asperger syndrome  
Autism describes symptoms seen  
together: social difficulties,  
communication difficulties, and  
unusual or repetitive behaviors.**

**Pervasive developmental disorders:  
autism, PDD, Asperger syndrome**


**Autism describes symptoms seen together: social & communication difficulties with unusual or repetitive behaviors.**

**It is not a separate disorder.**

**1 out of every 10 we can find the cause of the autism.**

**Causes are fragile X syndrome, Rett syndrome, chromosomal abnormalities, and SLOS.**

**Half of individuals with SLOS have been diagnosed with autism spectrum disorders.**



**Autism classrooms are smaller in size,  
emphasize visual instructions.  
You can use a picture board that shows  
the activities of the day.  
An autism school program usually has  
social skills training.**

**I also ask**

**1) what medical and psychiatric illnesses run in the family,**

**2) medical illnesses the child has – including allergies, sinus infections, ear infections, GE reflux, headaches and seizure disorder.**



**Is it because of physical illness?**

**Is it thyroid imbalance (can cause behavioral problems, mood and sleep problems but is not common). Thyroid stimulating hormone (TSH) is the blood test.**

**Is there  
attention deficit hyperactivity  
disorder (ADHD),  
mood disorder: bipolar disorder,  
disruptive mood dysregulation  
disorder,  
obsessive-compulsive disorder  
(OCD)?**



**I ask questions about:  
sleep, behavior, energy, appetite,  
moodiness, irritability, crying spells,  
concentration, self-injury (including  
skin injury, eye problems),  
aggression, “obsessions”, repeating  
behaviors.**

**Behavioral assessment: what is the purpose (the function) of the behavior if it is not a physical or mental health condition?**

**Is it for communication or for attention?**

**Is it for physical sensation?**

**Increase communication Sign language, picture exchange communication system (PECS).**

**Physical restraints can help self-injury & stiffness in arm sleeves can be decreased.**



**You can consider medicine if behavioral treatment isn't working and a child has one of the three criteria. The child is**

**1. suffering**

**2. a danger to herself or others**

**3. impaired in school or home life**

**Hitting the head can damage hearing by damaging the eardrum.**

**Hitting the head can also cause partial or total blindness by knocking the tissue on the back of the eye loose (retinal detachment)-ocular ultrasound can be performed**



**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 of using a medicine.**

# **Neuroleptics (antipsychotics):**

**treat aggression, disruptive  
behavior and self injury in autism  
and other developmental and  
genetic disorders**

**Risperdal (risperidone),  
Geodon (ziprasidone),  
Zyprexa (olanzapine)  
Seroquel (quetiapine),  
Latuda (lurasidone)  
Abilify (aripiprazole)  
Haldol (haloperidol)**





**Aripiprazole (Abilify) and haloperidol (Haldol) are not our first choice because they may increase 7DHC levels more than the other neuroleptics. But they might help.**

**Neuroleptics all can cause:**

**a temporary muscle stiffness (EPS)**

**a movement disorder that might not  
go away (Tardive Dyskinesia)**


**Neuroleptic Malignant Syndrome**

**You can test “blood levels” of neuroleptics such as Risperdal (risperidone) and Zyprexa (olanzapine) if the doctor is hesitant to go up on the dose of the medicine or the insurance company does not want to pay for a higher dose.**

## **Obsessions and compulsions:**

**An obsession is a thought that comes to you for you to do something when you don't want to do.**

**A compulsion it is the act of doing the obsession you're thinking of.**



**Compulsions: opening and closing doors and drawers, turning light switches on and off, emptying out bottles, repeating behaviors, throwing things away, touching things and collecting things.**

# Anxiety Disorders and Selective Mutism.

- Anxiety is often seen with SLOS,
- Often it is socially related anxiety
- Selective mutism is when a person can speak in some settings but does not speak in other settings (often speaking little at school).

**Antidepressants like Prozac are used for OCD and anxiety.**

**The SSRI Lexapro (escitalopram) is a good choice.**

**Be careful if there is a family history of bipolar disorder.**

**Cognitive behavioral therapy (CBT) can help;**

**Prozac (fluoxetine), Celexa (citalopram), Zoloft (sertraline), Paxil (paroxetine). Lexapro (escitalopram) is less likely to interfere with other medicines.**



**Benzodiazepines - Valium (diazepam), Ativan (lorazepam) can help anxiety and selective mutism (but are not often used in children) as a daily medicine.**

**Benzodiazepines capitalized not can also treat some seizure disorders.**

**Stressful or loud events: DVD or favorite movie via headphone, moving dose times around for medicines already being taken for behavioral problems (such as in afternoon med being given in the morning), Benadryl (but some people get worse).**

**Travel, medical procedures, blood draws, and emergency calming:  
Ativan (lorazepam); highest dose is usually 0.025 mg per kilogram (0.01 mg for every pound for each dose).  
Need at least 4 hours between doses.**

## **MOOD disorders:**

**Medicines for depression are antidepressants: Prozac [fluoxetine], Wellbutrin [bupropion], Celexa [citalopram], Lexapro [escitalopram], Zoloft [sertraline], Paxil [paroxetine]**

**For people who feel depressed in the winter (have seasonal depression) light boxes can be helpful (has side effect of sometimes worsening bipolar disorder).**

**Exercise can also be helpful for depression.**

**Bipolar disorder is an illness in which people can feel angry or irritable and at other times can be overly happy.**

**Many people with SLOS look like they have bipolar disorder. This is even when they don't have a family history of bipolar disorder**

**Medications for Bipolar disorder:  
lithium, Depakote (divalproax  
sodium), Depakene (valproic acid),  
Trileptal (oxcarbazepine), Tegretol  
(carbamazepine), Lamictal  
(lamotrigene).**

# Attention deficit hyperactivity disorder

-attention problems & hyperactivity  
or just attention problems.

-Can be looking & not attending well.

-Girls tend to have attention problems  
without as much high activity.

-inattention may be due to delayed  
processing time and not ADHD.



- Methylphenidate family (Ritalin, Metadate, Concerta, Focalin)**
- dextroamphetamine family (Dexedrine, Adderall [mixture of dexedrine salts], Vyvanse)**
- clonidine (Kapvay is long-acting)**
- guanfacine (Intuniv is long acting)**
- Strattera**

**For Blood tests, vaccines, procedures:  
Gebauer Pain Ease or other topical  
skin refrigerant spray can numb skin,  
Emla (lidocaine/prilocaine) cream can  
numb the skin of the arms but can  
make it harder to see the vein.  
Consume a lot of water in the a.m.**