

Behavioral Management in DESSH and other Neurodevelopmental Disorders

Paul E.A. Glaser MD, PhD
Department of Psychiatry
Division of Child Psychiatry
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Characteristic Behavioral Troubles in DESSH

- Anxiety Disorders
- ADHD (includes impulsivity-inattention-hyper)
- Aggression
- Autism Spectrum Disorder (or features of Autism)
- Mood Swings (now called DMDD)

What is the possible reason?

- Hannah C Rudolph et al. *Biology (Basel)*. 2023 Apr 12;12(4):589. Structure-Function of the Human WAC Protein in GABAergic Neurons: Towards an Understanding of Autosomal Dominant DeSanto-Shinawi Syndrome
- This would suggest that dysfunction in GABA neurons may lead to a hyperglutaminergic state in DESSH (Dr. Glaser's unproven hypothesis)

Applied Behavior analysis

- Rooted in behavioral techniques originally described by B.F. Skinner
- Should be overseen by Board-Certified Behavioral Analysts (BCBAs) and implemented by Registered Behavioral Therapists (RBTs)
 - Systematically observes, describes, and assesses behaviors
 - Works with the patient and family on finding reliable rewards
 - Devises plan to shape behaviors that lead to safer outcomes
- Wide range of evidence supporting its utility

Parent-Child Interaction Therapy

- Indicated for ADHD and oppositional behaviors/emotion dysregulation
- Parent training, typically “coached” by therapists who are observing through “bug in the ear”, video feedback sessions
- Works to establish warmth between parent and child, in addition to approaching problem behaviors and interactions
- Does require higher communication capacity than ABA

If Aggression Is Severe

- Atypical Antipsychotics
 - Risperdal (risperidone) FDA approved for Autism
 - Zyprexa (olanzapine)
 - Seroquel (sertraline)
 - Geodon (ziprasidone)
 - Abilify (aripiprazole) FDA approved for Autism
- Others
 - Naltrexone (Revia)
 - Lithium
 - Amantadine

If Anxiety/Mood Is a Problem?

- Try SSRI first
 - Zoloft (sertraline)
 - Prozac (fluoxetine)
 - Lexapro (escitalopram)
 - Celexa (citalopram)
 - Luvox (fluvoxamine)
- Others
 - Cymbalta (duloxetine)
 - Effexor (venlafaxine)
 - Buspar (buspirone)

If Impulsivity/Attention Is a Problem

- Stimulants (sometimes do not work)
 - Concerta, Metadate, Ritalin., Focalin (methylphenidate)
 - Adderall, Vyvanse (amphetamines)
- Clonidine (clonidine)
- Tenex (guanfacine), Intuniv
- Strattera (atomoxetine)
- Wellbutrin (bupropion)

- Memantine (Namenda)

Anti-Seizure Medications That Modulate Glutamate/GABA balance

- Lamictal (lamotrigine)
- Topomax (topiramate)
- Trileptal (oxcarbazepine)
- Depakote (divalproic acid)
- Tegretol (carbamazepine)
- Neurontin (gabapentin)

The Best Medications

- Structure
- Sameness
- Predictability
- Keeping Promises
- Simplicity
- Family Focus
- Consistent Discipline

Questions?

