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Reaching Out, Providing Help, Giving Hope

Management of Anxiety and Aggression

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People with Cornelia de Lange Syndrome (CdLS) and their families face special challenges. Children with CdLS often have cognitive and communication limitations, meaning they are at higher risk for developing behavior problems. There are interventions that may help prevent significant behavioral problems and there are treatments for the challenging behaviors that do develop. In general, it is important to understand a child's needs and to understand what causes the undesired behavior. Often this requires the help of a professional. A metaphor used by behavioral specialists at the University of North Carolina is that of an iceberg. The specific behaviors that we see in our children are the part of the iceberg that is visible above the water. The parts of the iceberg that are underwater are the underlying deficits or difficulties that lead to the behavior. They are the explanation for the behavior. Observing and collecting data about when an undesired behavior occurs can lead to an explanation of that behavior and to an intervention to prevent its recurrence.

Factors that Cause Anxiety and Aggression

There are many reasons for anxiety and aggression. General issues like change in routine, communication frustrations, social confusion, hormonal changes, and sleep changes can cause a change in behavior. Always make sure that there is no active medical issue that may be responsible for behavior changes. Manifestations of behavior problems can range from hitting and yelling, to destroying things, and even self-injury.

Prevention as a Strategy

Early intervention can help build life skills and can help circumvent behavioral problems in the future. Four general skill areas of focus are: **communication, education, activities of daily living, and social skills.**

1. Maximizing communication skills is critical in children with CdLS. Even if children are nonverbal, there are adaptive devices, sign language, picture cards, etc., that may be used to help them communicate. It is common for behavioral problems to develop in individuals who have limited ways to express themselves. Imagine how frustrated any of us would feel if we could not speak and had no way to communicate!
2. Developing basic academic skills is also critical. While many of our children have some form of mental retardation, they often can be taught basic reading and math. These skills will be invaluable to them as an adult.
3. Learning how to set the table, shop at the store, and do a load of laundry will prepare our children for adult life. There is obvious functional benefit in learning activities of daily living, and these skills increase self-esteem.
4. Finally, social skills may be difficult for a person with CdLS. They have trouble understanding social behavior and cues, and can have difficulty establishing



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friendships with peers. Some of these skills can be taught and will help our children succeed.

The Philosophy of Prevention Structure

Developing structure in our children's lives can do a lot to decrease anxiety. Children and adults with CdLS should have a daily schedule (with pictures or words) that allows them to know what activities are going to occur, when they will happen, and with whom they will occur. An example of a daily schedule is illustrated in figure 1. Imagine if you didn't have a calendar, daily written schedule, or planner. Wouldn't life seem confusing and unpredictable? The physical environment at home and school should be set up to minimize distractions and reduce stimulation.

Figure 1: Example Schedule

1. Eat breakfast
2. Get dressed
3. Brush teeth
4. Go to school
5. Eat snack
6. Toilet
7. Homework with Mom
8. Go for walk with Dad
9. Eat dinner
10. Jump on trampoline with Daddy
11. Go to bed

Exercise

Teaching our children different ways of exercising can help them to stay in shape and reduce their stress/anxiety level. Fast walking, swimming, and riding a bike are a few examples of activities that some children with CdLS can enjoy. Sometimes these activities can be used as a preventative measure to keep anxiety low and as an intervention to help kids "blow off steam." Programs like Special Olympics can help build confidence and foster peer relationships.

Calming Activities

It is important for our kids to know how to calm themselves. Teaching them skills ahead of time, i.e. "practicing leisure" is extremely useful for keeping anxiety down and for lowering anxiety/agitation when it is high. Examples of calming activities include listening to soft music, holding a special doll or stuffed animal, or lying in a hammock. Every child has specific activities that can work for him/her.

We can teach our children muscle relaxation by having them squeeze a ball, then release it. We can teach them deep breathing by demonstrating a deep breath and having them imitate. It is important that calming activities be part of the daily routine. They should be a



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part of the daily school schedule, along with reward activities (fun activities that motivate the child).

Intervention Strategies

Even with the best planning and parenting, behavioral issues can arise. It is important to develop a communication system that allows the child to tell you that they are upset or angry (just a picture of an angry or frustrated face that the child can point to will suffice). Create a place for the child to go when they are angry (beanbag chair, special chair, etc.). Add calming activities. As an example, in our house we have used “the red chair” as a place for Daniel to go when he is angry. Often he will sign “mad,” will need to be led to this chair, and will have 10 minutes of “quiet music” in order to calm down. These strategies allow for both parent and child to calm down and then afterwards, sort through the problem. Provide a clear beginning and end to the intervention (an egg timer is helpful). Also, develop alternative strategies in case the intervention does not work. Examples include pinching a beanbag, squeezing something, etc. It is better to use inanimate objects that can be pinched or squeezed than for children to pick an alternative, for example, their siblings.

Social Stories

These are stories that are written to help with a difficult situation that is anxiety provoking for the child. In the story, the child is the star. The story is about the event and is written so that the child is successful. This story is read multiple times to the child. The story can be put on picture cards, and the child can practice “telling” the story to you. Reading about anxiety-provoking events, such as flying in a plane or swimming, can lower anxiety, provide a sense of mastery, and allow the child to anticipate that the event will occur. Putting that activity on their schedule and letting them know when, where, and how the activity will occur should help. An example of a social story is given in Figure 2.

Figure 2. Example of a Social Story: Daniel goes swimming with Kristin

Every Wednesday, Daniel goes to the YMCA with Kristin. First, they go in Kristin’s gray car. Daniel likes to ride in Kristin’s car. They get to the YMCA and Daniel puts on his orange bathing suit. They get ready to go into the water. Daniel is scared, but Kristin helps him. Daniel gets into the water. He is happy because the water is warm. He has a lot of fun swimming. Good job, Daniel!

Examples of Specific Aggressive Behaviors and Interventions:

Pinching: give child a beanbag to pinch instead of other people

Biting: teach child to bite a banana instead of another person.

Destruction of books by ripping: Get laminated books. Allow them to shred scrap paper.

What to do if an Intervention Fails:

Sometimes, it takes multiple trials to figure out strategies that will work. Consider revising your intervention strategy. As your child matures, you may need to modify both the types of



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calming strategies and your interventions. Seek professional advice (school behavioral specialist, developmental psychologist, psychiatrist) if needed.

Medications

Prior to using medication, be sure that a medical problem is not causing the behavior. Make sure that your child has been tested for reflux and is being adequately treated for this condition. Before using medication, also look for environmental causes. Is there a pattern to the behavior? Does it happen at a certain time or place, or with the same person? Look for preventative strategies that may help decrease the frequency of the behavior. Think of interventions that can help your child control his/her challenging behavior (improved communication, calming activities, etc.). Prior to using medication, seek the help of a behavioral specialist.

There is a lot that we don't know about the use of psychiatric medication in the developmentally disabled. Few controlled clinical studies have been done. All classes of psychiatric medication have been used in children and adults with CdLS. It is somewhat "hit or miss" as to whether medications will work for your child's behavior problem. Medication treatment is complicated.

The following is a summary of general categories of medications that have been used in CdLS:

Antidepressants:

These medications are usually very safe and have few side effects. They are helpful for depression, agitation, anxiety, and obsessions/compulsions. **They take at least a month to begin working.**

Examples of antidepressants include: Prozac, Paxil, Zoloft, Luvox, Celexa, Lexapro. Remeron and Wellbutrin are other examples. Wellbutrin can also help for Attention Deficit Disorder (ADD).

Antipsychotics:

These are medications first developed for the treatment of schizophrenia. They are not specific for developmental disabilities, but can be used for aggressive/self-injurious behavior. The old antipsychotics include: Haldol, Prolixin, Navane, Mellaril. They are not used much anymore and can have long-term neurologic problems associated with them (called Tardive dyskinesia). Examples of the newer antipsychotics include: Zyprexa, Geodon, Risperdal, Seroquel, Abilify. There is an increased possibility for developing diabetes and high cholesterol with Zyprexa. It can also cause weight gain, which can be problematic in our children who may gain weight anyway at adolescence.

Other medications used to help with behavior include:

1. Anticonvulsants (seizure medications): Depakote, Tegretol, Lamictal, Gabitril, Gabapentin.
2. These are used for bipolar disorder, can level mood, and decrease irritability.



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3. Stimulants: used for attentional symptoms.
4. Opioid antagonists: Naltrexone, which has been used for self-injury that is difficult to treat.
5. Clonidine and Buspar are used for anxiety and aggression.

It is difficult to anticipate the behavioral problems that may arise in a child's life. Even the best parents struggle with these issues. Building life skills, providing a structured environment, ensuring regular exercise, and teaching calming activities will help to minimize the difficulties our children may face. Medication may also be helpful when these strategies do not work. Consultation with a specialist is often helpful.

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