

# Helping Your Child Cope with Childhood Cerebral Adrenoleukodystrophy (ALD)

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## *Making Functional Tasks Easier*

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This handout was written by physical therapists, occupational therapists and speech language pathologists at the University of Minnesota Masonic Children's Hospital. It includes information on ways that families can help their child with ALD cope with the most common symptoms of the disease. The symptoms are listed from most common to least common.

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## About ALD

Adrenoleukodystrophy (uh-dree-noh-loo-koh-dis-truh-fee), or ALD, is a genetic (hereditary) disease. In some cases, it damages the myelin, a thin covering that protects the nerve cells in the brain or spinal cord. These nerve cells tell your body how to think and move. When the myelin is damaged, the nerves stop working well over time. This can cause neurologic (brain) symptoms, like problems with hearing, vision, thinking and muscle control.

ALD has many forms. We can't predict which patient will get what form. Childhood cerebral ALD only affects boys. It causes myelin loss in the brain at a young age. It usually appears between ages 4 and 10. The myelin is more damaged over time. This means symptoms often get worse over time.

## Attention and impulsivity

The frontal lobes of the brain help control skills and behaviors. They help us start and stop actions, check our own behavior and change that behavior if needed. These lobes also help us plan future behavior for new tasks based on past experiences. They control basic functions like directing and keeping attention and controlling our response to our surroundings. In children with ALD, these frontal lobes can be damaged. This means your son may not be able to stop some actions, words, emotions or behaviors from happening. You may notice personality changes as well.

### *What does this mean for my child?*

- Your child may start to have trouble in school because he has trouble getting and staying focused in class.
- Your child may do things without thinking of the consequences. This can lead to safety concerns.
- Social skills may suffer because your child can't "filter" what he says and thinks.

- Since your child's brain doesn't check behavior, he may not act the right way in each situation.

### *How can I help my child?*

- Limit distractions in his surroundings. This includes clutter and background noise like TV and radio.
- Let him take breaks if he has focused for a long period of time.
- Use cues in his daily routine to remind him to stay on task. This may include picture cards or schedules, verbal reminders, touch and so forth.
- Break tasks into steps to help him better focus on each step.
- If your child also has vision or auditory processing problems, please see pages 3–4 for more ideas.

### *Who else can help?*

Ask your child's doctor if a referral to one of the following specialists may help your child.

- A **neuropsychologist** diagnoses, treats and helps patients manage brain-related problems and injuries. He or she can help your child cope with daily activities and school.
- An **occupational therapist (OT)** can help your child with daily living activities, managing emotions and coping at school.
- A **speech language pathologist** can help your child with communication, social and cognitive (thinking and memory) skills.

## Vision problems

A child with ALD may have changes in his vision. These changes may include cuts in the visual field, tunnel vision or total vision loss. A cut in a visual field is when the child can't see in a certain area, such as when looking to one side, looking up or looking down. Tunnel vision is when vision is reduced to a very small, central portion of what there is to see.

Vision problems with ALD are often caused by myelin loss in the parts of the brain that control vision. In other words, the eyes still work, but the brain can no longer tell what the eyes are seeing. For this reason, eyeglasses do not usually help with ALD vision loss.

### *How can I help my child?*

- Before asking your child to do a task, make sure that the task is in their visual field.
- Urge your child to turn his head when walking to scan for obstacles in his path.
  - Outline doorways and stairs with bright red or yellow tape to help your child see them.
  - Don't change the layout of your living spaces. Keep furniture in the same place. Reduce clutter in the room.
- Use your child's visual strengths. Give him visual information with high contrast (like black and white) and within the visual field that he can best see.
  - When setting the table, use a dark tray or placemat under a light colored plate so that the plate is easier to see because of the high contrast.
  - Use contrasting colors to make liquids easier to see. Put milk in a dark mug and grape juice in a white mug.
  - When brushing teeth, place the toothbrush on a washcloth that is a contrasting color.
  - Reduce clutter on the table and countertops.
- Try using large print for reading with a simple black and white format. Your therapist may suggest computer or iPad applications for communication and learning.
- For safety, use plastic instead of ceramic or glass that might break if dropped. Your child may not see the broken pieces.
- For more extreme vision problems, use touch to enhance what your child is able to see. For example:
  - Let him both see and feel the spoon before feeding him.
  - Use a magnetic board with raised letters so he can both see and feel the print.
  - You may need to physically guide or help your child complete a task. You may need to help your child put tasks in the right order, such as when getting dressed or brushing teeth.
  - If your child has both hearing and vision problems, loud computer games, TV or radio sounds in the room may make it harder for him to concentrate.

If your child has memory, thinking or hearing problems in addition to vision problems:

- Your child will need more time to process what he sees, hears or reads. Keep information short and simple. Don't give directions with a lot of steps. You may need to repeat what you said, or offer the information in a different way.

### *Who else can help?*

Ask your child's doctor if a referral to one of the following specialists may help your child.

- An **ophthalmologist** can give vision tests. They can also help with any treatments for vision loss not related to ALD.
- An **OT** can help your child learn how to best use remaining visual skills.

- A **low vision specialist or specialist for the blind** can help with adaptations to home or school environments.

## **Auditory processing**

Some children with ALD seem like they can't hear what is said to them. But this trouble is with auditory processing, not hearing. Auditory processing is how the brain knows and interprets sounds. It is not the same as hearing loss. The ears can still hear sounds, but the brain can't tell what it is hearing. This is true for all sounds in the child's environment, including speech. Some children with ALD may have changes in auditory processing skills.

### ***What does this mean for my child?***

- It is harder to process sounds when it is noisy, as your child needs to listen to what you say while also tuning out background noise.
- Some boys have trouble controlling how loudly they speak. This is because the brain doesn't properly send a message about how loudly or softly he is speaking.
- Your child may have trouble following directions. He may not understand what you are asking.

### ***How can I help my child?***

- Avoid background noise when you talk with your child.
- Turn the TV off when you ask your child to do something.
- Speak slowly and in short sentences.
- Allow extra time for your child to figure out what you said before asking again.
- Don't speak louder. The ears can still hear a whisper or a shout, but the brain isn't sure of the message.

- Don't repeat information over and over again in a short time.
- Visual or touch cues can help. Show your child what you want or help him touch the object he needs to use. If your child is able to read, use a whiteboard and wide, dark marker to write down what you want your child to do.
- Have your child restate what you said to make sure he understood you.

### ***Who else can help?***

Ask your child's doctor if a referral to one of the following specialists may help your child.

- A **speech language pathologist** can help your child with communication issues.
- An **OT** can help with adaptations at home and school to make up for gaps in understanding.

## **Communication**

It can be hard to talk with a child who has ALD. Many children with ALD have communication issues. These can be any combination of the following:

- trouble understanding speech and language (auditory processing)
- memory problems
- trouble making sounds and words (motor ability)
- trouble forming what the child wants to say, or not wanting to speak

### ***How can I help my child?***

- Give your child extra time to respond to what is said.
- Speak in short, simple sentences. Give your child a chance to respond. If there is no response, repeat what you said.

- If the first 2 items don't work, try asking your question so that it can be answered with a yes or no.
- Accept all types of communication. Your child may speak, make a sound, gesture, point or write. Fatigue (tiredness) and medicines may cause your child's ability to communicate to differ during the day.
- Make sure you look at your child when you speak to him. Facial expressions add meaning.
- Some children with ALD are able to read and write. They may respond well to simple words written on paper or a dry erase board.

## **Motor planning**

Motor planning (praxis) is the process of deciding what your body has to do and then doing it. It is a very complex function and involves many parts of the brain. Motor planning includes thinking of, planning, sequencing and doing actions. Children with ALD may have trouble planning big body movements or fine motor (hand) movements.

### ***What does it mean for my child?***

Trouble with motor planning looks like clumsiness or lack of coordination. Your child may:

- Have trouble doing simple tasks he already knows, such as getting dressed or walking to the bathroom.
- Sound like he is slurring his words and may be hard to understand. This is because motor planning problems can involve the muscles and nerves that move the lips, tongue and jaw.
- Get easily frustrated and avoid motor activities.
- Have trouble swallowing or managing his spit (saliva).

### ***How can I help my child?***

- **Be patient.** Your child is likely frustrated too.
- Expect that teaching new skills will take a lot of time, practice and repetition.
- Touch can improve body awareness. Try deep lotion massages and regular range of motion exercises for each joint of the body.
- Use touch to help your son with certain parts of a task. Using your hands to guide his hands is a great way to help him learn or relearn a motor task.
- Activities that include mimicking motor movements are helpful. Examples include playing "Simon says" or copying new dance moves or actions to songs.
- Children with motor planning problems often also have visual or auditory processing problems. Please see pages 3–4 for more tips.

### ***Who else can help?***

- Physical, occupational and speech therapists can help you and your child learn how to cope with motor planning problems.

## ***Dysphagia***

Dysphagia means having trouble eating or swallowing. ALD can affect the nerves that control eating and swallowing. Eating problems include:

- trouble chewing or chewing too much
- being picky about types of food
- choosing soft or even pureed foods

Swallowing problems include:

- trouble starting a swallow
- coughing, choking or turning blue after a swallow

### ***What are the symptoms?***

The most common symptoms are:

- Holding food in cheeks
- Taking longer to eat
- Fever spikes
- Pneumonia
- Poor appetite

More subtle symptoms include:

- Weight loss despite eating
- Chronic cough
- Upper airway congestion
- Lethargy
- Drooling
- Decreased hunger
- No chewing

### ***What does it mean for my child?***

Dysphagia can lead to poor nutrition. Poor nutrition can lead to low energy, weight loss and medicines not working as well. The disease can also put your child at a greater risk for infections, breathing troubles and aspiration pneumonia (food and drink in the lungs).

### ***How can I help my child?***

- Change your son's diet to foods he will tolerate. This could be soft solids, pureed food, strong flavors or hot or cold temps.
- Don't push or nag your child to eat. Instead of 3 larger meals, try offering small snacks 5 to 6 times during the day.
- Give your child extra time to eat. This lets your child manage the food in his mouth and feel more ready to swallow.
- Some boys will need a feeding tube for extra nutrition. The tube may be used part time or all the time. Your doctor will talk to you about the best option for your son.

### ***Who else can help?***

- **Speech pathologists** can help you and your child learn how to eat safely. They will also teach you about risks of eating when the swallow changes.
- A **video swallow study** may also be helpful. This can tell us more about why your child is having trouble swallowing.



## Muscle Tone

Children with ALD often have too much muscle tone (hypertonia). Muscle tone and muscle strength are not the same thing. Muscle tone is the tension (pressure) between the muscle fibers. Muscle strength is the power the muscle fibers can create. Extra muscle tone limits how much or how easily a muscle can move. It is sometimes painful and can lead to limited muscle movement (contracture), deformed joints or total loss of muscle function.

### ***What causes it?***

Muscle tone is controlled by brain signals that tell nerves to move the muscles. When the part of the brain in charge of these signals is damaged, it causes hypertonia.

### ***What does it look like?***

Hypertonia is most often first seen in the calf muscles. Some children will have hypertonia in their whole body. Symptoms to look for are:

- Stiff muscles that make it hard to bend or straighten a joint
- Arms and legs that are bent or straight without your child's control
- Clonus. This is a shaking movement, most often seen in the hands and feet. The muscle will quickly move between contracting and relaxing. It is most often seen if a muscle is fatigued or if the limb is moved quickly.

### ***How is it treated?***

- **Physical and occupational therapy.** We will show you and your child how to help relax the muscles. This includes range of motion exercises, stretches and positioning tips. These therapies can increase your child's comfort. It can also help reduce loss of function caused by the joints not moving fully. Your therapist may also suggest positioning aids and equipment to help treat muscle tone.
- **Medicines.** Your child's doctor may give your child medicines to help with symptoms.

## Mobility and positioning aids

**Joint braces (orthotics) or splints.** These can help the joints align better, which reduces muscle tone.

- Splints are often used when the child is resting. They are often worn at night on the hands or the feet.
- Braces are most often used on the feet to help your child get around.
- If your child has tight heel cords (Achilles tendon) and some walking trouble, he may need ankle or foot braces. These will help relax the muscle tone and prevent more shortening of the heel cord. They will also help with balance and muscle tone while walking.
- An occupational or physical therapist can help decide if these are right for your child.

**Mobility aids.** Some children with ALD may have trouble with sitting balance and walking. Mobility and positioning aids are tools that help these children be active at home, school and in the community. These aids may include car seats, strollers and wheelchairs.

## **Car seats**

These are needed for safety if your child can't stay sitting on his own in a moving vehicle. You can buy car seats that will fit children who weigh up to about 100 pounds (45.36 kg).

## **Medical strollers**

These fit children of all sizes up to adult. If a child does not have high muscle tone and has fair sitting balance, you can use a stroller for long distances. Strollers are for mobility (getting around) only. They should not be used for long periods of sitting.

## **Wheelchairs**

This is the best option if your child:

- Has high muscle tone
- Has problems with sitting balance
- Can't walk without a lot of help

Wheelchairs differ from strollers because you can adjust them so that they “grow” with the child. Wheelchairs can also be used with seating and positioning inserts for children who have high muscle tone and trouble with sitting balance. You can use wheelchairs for mobility and for long periods of sitting.

## **How do I get mobility and positioning aids?**

- Your physical or occupational therapist will help you choose the right aids. They will teach you and your child how to use the aids safely. Your doctor will then write a prescription for it.
- When you will get the aid depends on your insurance company or other payment source. It may also depend on the company that makes the aid if you are ordering a custom piece.
- You may not be able to get custom aids until your child is in outpatient therapy in your home state. Custom aids need to be approved by a doctor before you buy, and they take time to build. You may rent aids while you wait.
- Your insurance company may ask you to use an aid company in your state. Your therapist and discharge planner will help you plan this.