

**An Educator's
Guide to MCADD**

What is MCADD?

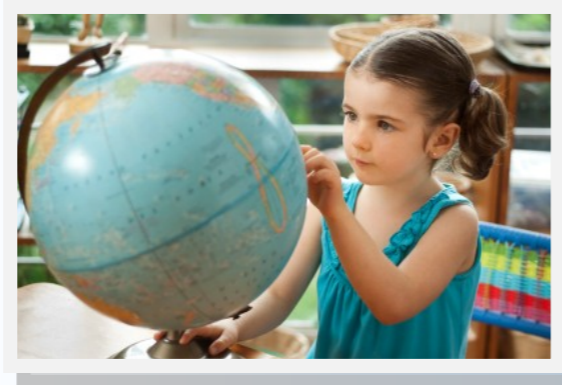
Medium chain acyl-CoA dehydrogenase deficiency (MCADD)

Background

MCADD stands for "medium chain acyl-CoA dehydrogenase deficiency". It is one type of fatty acid oxidation disorder that is inherited if both parents carry the gene for MCADD. Individuals with this type of disorder have problems breaking down fat into energy for the body.

In MCADD, the enzyme that breaks down certain fats in the food we eat is either missing or not working properly. Because of this, the body cannot use fat for energy and must rely only on glucose, a type of sugar. Once the glucose is used up, the body tries to use fat without success.

When we are ill, our bodies need extra energy to fight illness. Since children with MCADD cannot create this needed energy from fat stores, they are at risk for a metabolic crisis. MCADD is now identified through newborn screening which allows for early treatment.



First Signs of MCADD

Children with MCADD usually have symptoms for the first time between three months and two years of age. These symptoms are often brought on when a child stops eating, usually because of a common illness such as the flu, an ear infection, or a cold.

Metabolic Crisis

MCADD can cause a metabolic crisis. A metabolic crisis may start with symptoms of fever, diarrhea, and vomiting. This is usually followed by low blood sugar. If a metabolic crisis is not treated, it can lead to breathing problems, coma, or death. Prompt treatment can prevent these consequences.

Hypoglycemia

If children with MCADD go too long without food, low blood sugar (hypoglycemia) can occur, with or without other symptoms of a metabolic crisis. Low blood sugar can cause weakness, shakiness, dizziness, or cold, clammy skin.

Symptoms and Treatment

Common Symptoms

Symptoms often occur after a child has nothing to eat for more than a few hours. During long periods without eating food, the glucose in the body is used up. Some of the first symptoms include:

- Sleepiness
- Behavior changes
- Irritable mood
- Poor appetite
- Low Energy
- Fatigue
- Vomiting

Preventing Symptoms

When symptoms occur, a high carbohydrate drink or starchy snack may help. To prevent symptoms from occurring, children with MCADD may need to eat or drink more often than their classmates.

Children who are sick often do not want to eat, or they vomit and cannot keep food down. If this happens, their parents need to be called immediately, and the child may need to go to the emergency room for Intravenous (IV) treatment.



Dan Poel (Ohio Sports Imaging)

Treatments*

Treatments can include:

- Avoidance of going a long period of time without food.
- A low fat, high carbohydrate diet (some parents try to keep the child's fat intake to 25-30%).
- Prescription L-carnitine supplements that help the body make energy and break down fat.
- Cornstarch mixed in liquid can also help to decrease the frequency of morning hypoglycemia.
- IV treatment in the emergency room if the child vomits or refuses to eat.

*Note: Treatments will vary depending on the child's specific situation. Talk to the parent to see what is best for the child.

The Latest Research on MCADD

In a recent study at Children's Hospital Boston, the following information was obtained through questionnaires and interviews with parents who had children with MCADD.

Early Intervention & Motor Skills

- One-third of the children in the study received early intervention services for language delay.
- Some children with MCADD may have delayed motor skills in early development, but will catch up with the rest of their peers.
- Most children with MCADD will not have academic problems in school.

Internalizing Behaviors

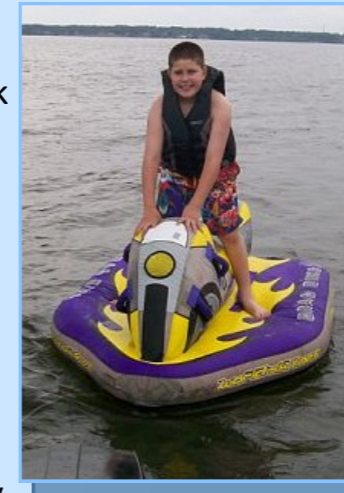
Over half of the children in this study tended to internalize their behaviors more than their peers. Children who internalize behaviors may:

- Monitor their own actions to excess
- Be more compliant, so their problems may go unnoticed
- Worry excessively (e.g. Say "I'm not good at this")
- Be very sensitive
- Have high anxiety
- Be more likely to withdraw from social situations

Narratives from Parents

Parent interviews took place to assess how the child was doing in school.

One parent commented that her child only received an extra snack if he was willing to ask the teacher for one. Because of this, he sometimes went without a snack and was very tired by the end of the school day.



Another parent stated that her son always had a hard time expressing his feelings.

"I always encouraged him to join activities, but it wasn't until he joined the school robotics team and had to make a presentation that he started to come out of his shell."

Results from this study indicated that children with MCADD need a little extra encouragement to join activities, speak up in class, or tell the teacher if they have a problem.

How Can Teachers Help?

Parents Recommend*

Parents asked that teachers inform them if:

- There is sickness going around school that might place their child at risk.
- A special snack will be served that is higher in fat than the child should eat (e.g. cupcakes for a birthday party).
- Their child is unable to focus in school, seems overly fatigued, or is acting different than usual.
- The class will be outside in hot weather for a long period of time (Because energy is required to keep cool, a child with MCADD may need to sit in the shade or have extra nutrients to maintain energy levels).

*Note: These recommendations may not apply to all children with MCADD. If teachers have questions on specific concerns, it is best to communicate these with the parent.



If there is a problem...

The child should see a nurse if he or she is experiencing extreme fatigue or other symptoms of illness.

The child may look and feel fine one minute and then have a severe problem the next. To reduce the chance of symptoms, encourage the child to:

- Stay well hydrated
- Always eat during snack time
- Tell the teacher if he or she feels ill
- Monitor how they feel in hot weather or after physical activity

The teacher should watch for signs that might indicate a problem.

Tips to Remember

Dietary Information

Children with MCADD may need more nutrients than their peers to maintain their energy levels and function in school.

Mothers recommend a mid-morning and/or mid-afternoon snack such as:

- Gatorade, Powerade, or Apple juice
- Popsicles
- Pudding
- Granola bars
- Low-fat cheese and crackers

One parent stated that highly processed, greasy, and fast foods were particularly difficult for her child to digest. To compensate for school lunches, she sent her child with healthy snacks to help him maintain his energy.

Another mother found that if her child was unusually irritable or tired, she would give him a high carbohydrate snack. Within about 15 minutes, the child would regain his energy and feel like his usual self again.



Children with MCADD may be fine one minute, and then be in need of serious medical attention the next. Even though they look healthy, the MCADD is still there.”

- Mother of a 7 year old with MCADD

In Conclusion

Most children with MCADD perform very well in school, but might need encouragement in the classroom and extra nutrients throughout the day.

Depending on the severity of the child's MCADD, the child may not have any symptoms. It is important to talk to the parent to know what to look for in case the child needs assistance.

Parents want the teacher to treat their child the same as all the other students, but also to realize that MCADD can be very serious. If the teacher has any concerns, these should be communicated with the parent to prevent future problems.

Resources & Notes

Resources

The New England Consortium of Metabolic Programs

This website contains educational materials and links to other resources. There are teacher resources located in the “For Professionals” section.

<http://newenglandconsortium.org/>

FOD (Fatty Oxidation Disorders) Family Support Group

The FOD Support Group provides emotional and grief support, practical information, and medical updates in screening, diagnosis, research and treatment.

<http://www.fodsupport.org/>

The National Newborn Screening and Genetics Resource Center (NNSGRC)

NNSGRC provides information and resources in the area of newborn screening and genetics.

<http://genes-r-us.uthscsa.edu/>

Teacher's Notes:



Notes & Contact Information

Notes from Parents:

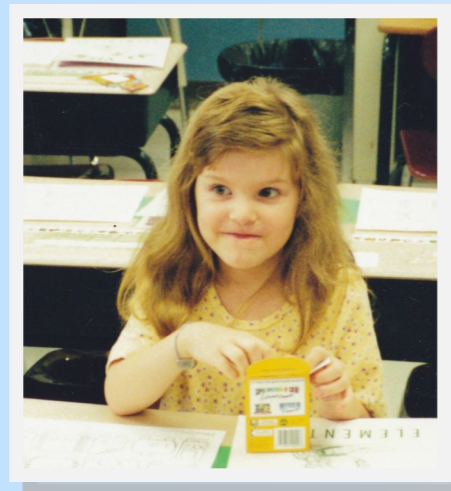
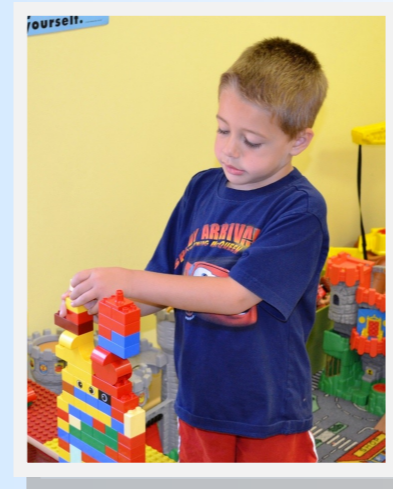
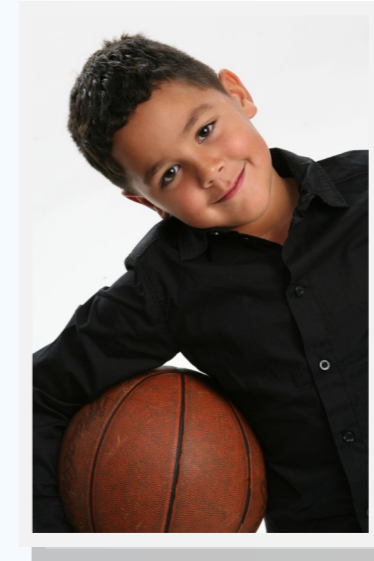
Parent Contact Information:

Physician Contact Information:

Acknowledgements

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Children's Hospital Boston



Created by Jenna C. Wilson, M.A.
Emerson College, Health Communication Program
Supervisor: Timothy Edgar, PhD
Preceptor: Susan Waisbren, PhD

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For specific questions, contact Susan Waisbren, PhD.
susan.waisbren@childrens.harvard.edu
Metabolism Program, Division of Genetics
Children's Hospital Boston, Fegan 10
300 Longwood Ave.
Boston, MA 02115