

to

**METHYLMALONIC
ACIDEMIA**

METHYLMALONIC ACIDEMIA (MMA)

These are the facts...

Methylmalonic Acidemia (MMA) is a rare inherited disorder of protein catabolism. Children with MMA do not have a functioning enzyme within their cells to metabolize or break-down four amino acids called isoleucine (ILE), valine (VAL), threonine (THR) and methionine (MET). These amino acids are found in all food proteins.



Protein in foods is important for building and repairing the body's tissues. Amino acids are often called the "building blocks" of protein. Twenty-two amino acids can be joined together in various combinations to form all the different kinds of protein in foods. Enzymes are special substances in the body which work to separate the amino acids in food proteins and recombine them to form different proteins in the body.

All children need a certain amount of ILE, VAL, THR, and MET for normal growth and tissue repair. In most people, any extra of these four amino acids is broken down and eventually used by the body in different ways. But, with MMA, extra ILE, VAL, THR and MET produces byproducts containing methylmalonic acid which are not broken down and can build up in the blood, spinal fluid and urine. Left untreated, these excess amino acids can interfere with normal brain development causing mental retardation and other life-threatening complications.



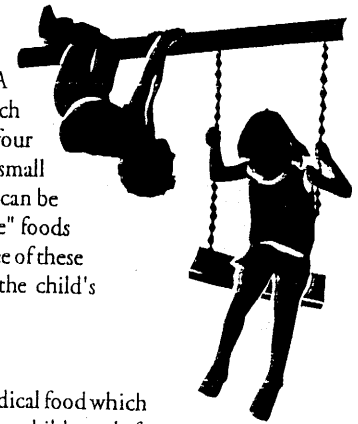
The good part is...

While there is no newborn screening program to check all babies for MMA, a baby with MMA may exhibit symptoms which lead to diagnosis within the first few months of life. Some children present with milder symptoms, and may not be diagnosed until they are older. The infant or child with MMA can then be put on a carefully controlled diet which allows just enough ILE, VAL, THR and MET for growth. Monitoring of these four amino acids with dietary adjustments is then done on a regular basis. Early diagnosis and continued dietary treatment are essential for normal growth and development to proceed in a child with MMA.

needs updating!

And now the diet...

Since ILE, VAL, THR and MET are found in all foods that contain protein, children with MMA must control their intake of protein. Foods which contain high amounts of protein are high in these four amino acids and should not be eaten. Foods with small amounts of protein and ILE, VAL, THR and MET can be eaten only in limited amounts. Some foods are "free" foods because they contain no protein and are therefore free of these four amino acids. They are eaten to help boost the child's intake of calories needed for energy.



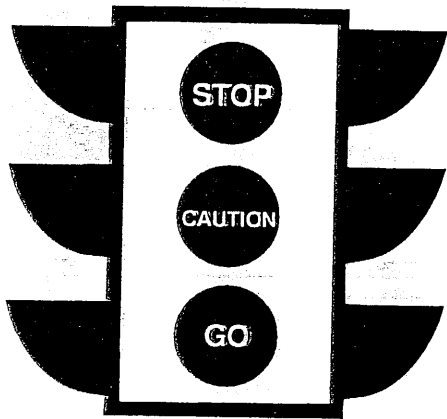
A child with MMA has a special drink called a medical food which has most of the protein, vitamins and minerals that a child needs for growth with little or no ILE, VAL, THR and MET. The special drink provides almost all of the nutrients that other children get from their food. It has a taste and smell that may seem objectionable to someone who is not used to it. However, children with MMA eventually acquire a taste for it.

The amount of drink and food the child consumes in a day is carefully calculated by the child's family, doctor and nutritionist. All foods must be carefully measured to control the amount of ILE, VAL, THR and MET that the child eats. These amounts are adjusted to the child's changing needs as he or she grows. A child with MMA learns at an early age that his or her diet is restricted and to ask a parent if a new food is allowed. Some children are given vitamin B₁₂ because their type of MMA is controlled by taking large doses of this vitamin. These children usually do not require a special diet.



Parents have...

extensive information on foods to use and avoid in feeding their child with MMA. If needed, they can provide a detailed list of foods with specific amounts that their child could eat. Some examples:



NOT ALLOWED...

meats, fish, chicken, turkey, milk, cheese, ice cream, yogurt, eggs, beans, nuts, peanut butter.

ALLOWED BUT CONTROLLED...

fruits, fruit juices, vegetables, vegetable juices, cereals, crackers, pastas, rice, potatoes-including potato chips, popcorn, some breads, special low protein foods. *

FREE FOOD...

sodas, Kool-Aid, lemonade, popsicles, jellies, gum drops, suckers, hard candies.

* Some food manufacturers use special ingredients to make low protein foods, such as pastas and baked goods, for this type of diet. Some of these items may be included as part of the child's meal or snack. These foods are an important addition for some children because they allow added variety without providing too much ILE, VAL, THR and MET.

How you can help...

Treat the child with MMA as a strong, healthy person. However, be aware that common childhood illnesses can be more serious for the child who has MMA and may lead to elevated blood levels of methylmalonic acid and related substances. Be alert to special learning needs, but remember that none may exist in this particular child.



Keep an open line...

of communication with the child's parents with notes or phone calls to reinforce parent teaching and monitoring of food eaten away from home. The child's parents are your closest experts on MMA.

Let the parent know...

- If the child has eaten any foods that are not allowed.
- If the child does not eat foods that are sent from home.
- If the child trades food with classmates.
- If the child's health or behavior seems markedly different.
- If special occasions such as birthday or holiday parties are planned so they can make sure there is a low protein food for the child. Parents may want to send a supply of low protein treats that store well and can be available for special or unexpected occasions.

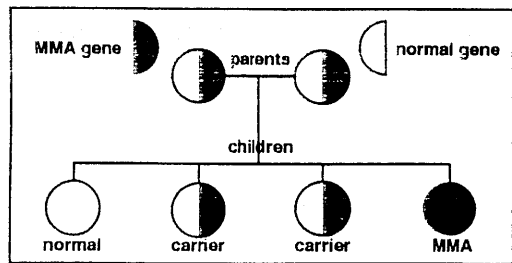
Please do not feed the child with MMA any foods that are not allowed on the previous list (on pg. 4) or lists provided by parents. Even "little tastes" add up and result in an elevation of methylmalonic acid and its byproducts in the child's blood. While the child may be curious about these foods, he or she is accustomed to doing without them.

Questions & Answers

Q How often are children with MMA born?

A MMA is a relatively rare disorder, 1 in 50,000 births. Different forms of MMA differ in severity, but all require specialized treatment. Treatment can be so effective that the children grow and develop normally and attend regular schools. Some children who are diagnosed and treated later may suffer from permanent neurological damage, causing mental retardation, seizures, poor muscle control and delayed development. Even with treatment, an infant or older child with MMA could develop complications that could lead to death.

Q Since MMA is inherited, do all the children in that family have MMA?

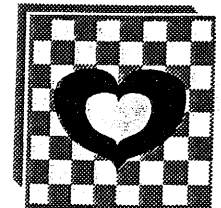
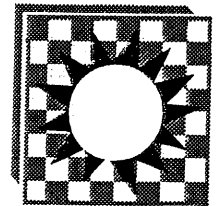


A For a child to have MMA, each of the parents must be a "carrier" of the MMA gene. A carrier has one normal gene and one MMA gene, but can break down ILE, VAL, THR and MET effectively. Carriers do not have MMA. A child with MMA inherited a MMA gene from each parent. When parents who are carriers have children, there is a 25% chance that the baby will be free of the MMA gene, 50% chance that the baby will be a carrier, and a 25% chance that the baby will have MMA. With each pregnancy, there is the same 25% chance that the newborn will have MMA. In some families, there may be only one child with MMA, while in other families, multiple children may be affected.

Q How do the parents know what to give the child with MMA to eat and drink each day?

A The child's diet depends on the individual child's diet prescription and food preferences. Diet calculations are based on one amino acid, valine. When valine is limited appropriately, ILE, THR, and MET will also be under control. The diet prescription tells how much special drink the child should have and how many milligrams of valine the child should get from table foods each day. Here is an example of a day's food intake for a 6 year old boy with MMA. His diet calls for a drink powder made especially for people who have MMA and 900 milligrams of valine from table foods.

	valine (mg)
BREAKFAST	
Rice Krispies, 1/2 cup	62
Non-dairy creamer, 1/2	96
Banana, 1/2 cup sliced	36
Orange juice, 1/2 cup	11
Special drink, 8 oz.	0
LUNCH	
Vegetarian vegetable soup, 3/4 cup	126
Saltine crackers, 6 squares	70
Grapes, 1 cup	30
Special drink, 8 oz.	0
SNACKS	
Popsicle, 1 stick	0
Raisins, 1/2 cup	48
Kool-Aid, 8 oz.	0
Graham crackers, 4 squares	96
DINNER	
Spanish rice (meatless), 1/2 cup	119
Green beans, 1/2 cup	48
Fruit cocktail, 1/2 cup	20
Ritz crackers, 12 pieces	132
Margarine, 1 pat	3
Special drink, 8 oz.	0
TOTAL	897



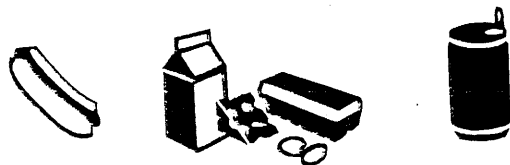
Q How can a child with MMA grow without high protein foods?

A The MMA child's special drink contains most of the protein, vitamins and minerals needed for growth. It really is a special drink! The food that the child eats provides the rest of the necessary nutrients. "Free" foods provide additional calories and variety.



Q If a child with MMA eats a high protein food, will he or she feel sick?

A Exceeding the daily limit of ILE, VAL, THR, and MET by eating a forbidden high protein food can disrupt the child's delicate biochemical balance. A serious medical emergency called ketoacidosis can result and may require hospitalization. Initial symptoms that may accompany a loss of biochemical control include a loss of appetite, vomiting, irritability, lethargy, and a change in muscular control. Also dangerous to the child is going without food for extended periods of time. This causes the child's body to break down stored proteins which will be used for energy, and releases ILE, VAL, THR, and MET into the blood.



Q Is the child with MMA sick more than other children?

A The child with MMA can be as healthy as other children. However, even initially mild illnesses can disrupt the MMA child's biochemical control and become life threatening due to complications. For this reason, it is important that the child with MMA receive appropriate immunizations and prompt medical attention if there is any sign of illness. Parents should be notified if classmates contract any contagious illnesses (like chicken pox) so that they can watch for symptoms in their own child and be prepared.



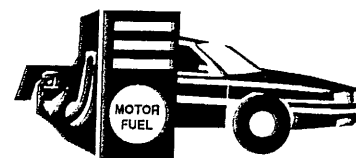
Q If there is a suspicion that the child with MMA is becoming sick due to illness or excessive protein intake, what should be done?

A Immediately alert the parents of the child's symptoms. Do not wait until the end of the school day. Offer water, Kool-Aid, clear soda pop, or Gatorade, if desired.

Q How can I explain the MMA diet to other children?

A Children can understand that since cars with different engines use different fuel (gas, diesel, etc.), some children have bodies that work in different ways and need different food. It is important that classmates do not trade food items or lunches with the child who has MMA.

Don't hide the fact that the MMA child's dietary needs are different. Speak with the child and/or the parents privately to determine how to answer in the best way.



Q When can the diet be discontinued?

A A child with MMA will never be able to metabolize ILE, VAL, THR and MET in the normal way. The risks for abnormal brain development and mental retardation become less severe as the brain matures and develops with age. However, the risks for ketoacidosis and neurological damage are always present. Continued diet therapy is essential.