To Request More Information:

Please answer the following questions and mail to Deb at the address below.

I am a:

- ☐ FOD Family
- ☐ FOD Familymember
- ☐ Professional

Check all that apply:

- ☐ I/my child has an FOD
- ☐ Please add me to your Mailing List Only
- ☐ Please add me to the printed Family or Professional List for networking purposes

Comments:

Name and/or Professional Title

Address

Phone [Deb calls new Families in the US and abroad]

Fax

Email

Please mail to:

FOD Family Support Group
Deb Lee Gould, MEd, Director
2041 Tomahawk
Okemos, MI 48864

The FOD Family Support Group is a family-based Support Group and a 501c3 (tax-exempt status pending) Non-Profit Corporation. Donations will be considered tax deductible later this year when we attain full tax-exempt status and will be retroactive to Jan 1, 2007. Your donation will help us cover copying, postage, website fees, phone calls to US and abroad, and Conference expenses, as well as Clinical/Research Funds. No donations are used for administrative salaries. If you write a US check, please make it out to the 'FOD Family Support Group' and mail to the address above. We also have a PayPal donation link on our homepage. You can designate your donation to the General FOD Fund (daily operations), FOD Clinical Fund, or the FOD Research Fund. THANK YOU from ALL of our Families!

For Families Living with a

Fatty Oxidation Disorder

FOD™

All in This Together

Fatty Oxidation Disorders
Family Support Group
www.fodsupport.org

Deb Lee Gould, MEd, Director
MCAD Parent and Grief Consultant
2041 Tomahawk
Okemos, MI 48864

Phone: (517) 381–1940
US/Canada Fax: (866) 290-5206
International Fax: (313) 432-5928
Email: deb@fodsupport.org
Website: www.fodsupport.org

501c3 Non-Profit Corporation (tax-exempt status pending)
Federal Tax ID # 83-0471342

FOD Family Support Group

Our History and Mission

The Fatty Oxidation Disorders (FOD) Family Support Group was 'born' in 1991 as a way of dealing with the sudden death of our 21-month-old daughter, Kristen, in 1985 from undiagnosed MCAD. Initially she was 'diagnosed' with Reye's Syndrome and it was not until one year later, when our son, Kevin, was born and diagnosed with MCAD that we discovered Kristen also had this rare metabolic disorder. Our third child, Brian, is a carrier and not directly affected. To say the least, our Family has been changed forever by this genetic disorder.

Our hearts
give immortality to those
we love in memory...

Our Mission is clear...to connect and network with FOD Families and Professionals around the world. Through our online newsletters, website, and Email List, we provide emotional support, practical information about living with these disorders, and inform Families of new developments in screening, diagnosis, treatment, and research.

Awareness of these disorders is imperative for early diagnosis and treatment, prompting us to be committed advocates for Expanded Universal and Comprehensive Newborn Screening for FODs and several other metabolic disorders. We do not want other Families to needlessly go through what we experienced.

From our Family to yours, we want you to know...

You are NOT alone
and
'We Are All in This Together'

Take care...
Deb and Dan
Kristen, Kevin and Brian Gould

FODs Defined

Fatty Oxidation Disorders are genetic metabolic deficiencies in which the body is unable to oxidize (breakdown) fatty acids to make energy because an enzyme is either missing or not working correctly. The main source of energy for the body is a sugar called glucose. Normally when the glucose runs out, fat is broken down into energy. However, that energy is not readily available to a child or adult with an FOD. If undiagnosed and untreated, these disorders can lead to serious complications affecting the liver, heart, eyes, and general muscle development, and possibly death. These disorders are sometimes misdiagnosed as SIDS and Reye's Syndrome.

Symptoms

There is a wide variation in the presentation of FODs, even within the same Family. Not every individual responds to a disorder in the same way. Some may be without major symptoms, yet others may have chronic bouts of low blood sugar or illnesses leading to hospitalizations. [Please note that blood sugar levels should not be totally relied on as the only indicator of a possible crisis - the levels may even look normal. However, some of our children/adults are symptomatic with glucose levels of even 70 or 80. Changes in behavior, irritability, lethargy (sleepy), and blood levels such as liver enzymes, acylcarnitines, ammonia, and ck (cpk) should also be monitored.]

An emergency situation exists when a 'metabolic crisis' occurs. A crisis is often preceded by a period of fasting, possibly due to vomiting or an infection. Other symptoms may include diarrhea, lethargy, seizures, coma and difficulty breathing.

Awareness of how to diagnose and treat these disorders is vital, because during a metabolic crisis, an undiagnosed individual may experience excessive buildup of fat in the liver, heart and kidneys, along with some brain swelling... all of which can lead to death.

Diagnosis and Treatment

Fatty Oxidation Disorders are autosomal disorders affecting both males and females. Both parents have to be at least a carrier of an abnormal gene (we do have parents WITH the disorder as well), in that when two abnormal genes unite the child will have an FOD. There is a 25% chance that EACH child will have an FOD and a 75% chance of being either a carrier or not having the disorder at all. If one child is diagnosed with an FOD, their siblings should also be tested, even if they are asymptomatic.

These disorders are diagnosed in a variety of ways at specialized Labs across the country. Blood, urine, skin fibroblasts, amniocytes (from amniotic fluid), and muscle and liver tissue are some of the specimens analyzed. The diagnostic tests often include an acylcarnitine profile, urine organic acid analysis, carnitine levels, and enzyme assays in fibroblasts. The acylcarnitine profile with whole blood on a 'PKU card' (filtered paper card) is the most direct approach for diagnosis of most of the FODs. Molecular DNA testing is also available for some FODs.

Treatment for FODs is multidimensional. A major concern is to avoid going without food for beyond 8-12 hours (i.e., an overnight sleep for a child or adult, when well). It is strongly recommended, however, for infants under age 1, that they should be fed around the clock every 2-4 hours. Most older children/adults with an FOD eat often throughout their waking hours (i.e., every 3-6 hrs). The interval may depend on various factors and their specific circumstances, such as if they are ill versus well, in which case they most likely would try to eat more often around the clock. Please note that fasting times can vary from person to person within the different disorders. A fasting state, especially while ill, can trigger a 'metabolic crisis' leading to lethargy and hospitalization. If hospitalized, it is imperative, according to FOD experts, that a 10% dextrose IV is started immediately following blood chemistry samplings. Waiting hours for the lab test results before putting in the IV can be fatal when an FOD child/adult is in crisis. [Note: Experts also recommend avoiding aspirin, oral products that contain salicylates, some steroid medications, and fat binding anesthesia medication].

Several snacks and meals of lowfat/high carbohydrate foods such as pasta, cereal, and other high complex carbohydrates (as well as sugar drinks when not well) are recommended throughout the day. Additionally, some Families use various lowfat nutritional supplements and MCT Oil (not used for all FODs, i.e., MCAD, since the medium chain triglycerides cannot be broken down). Infants under 10 months to one year should continue to have at least one night feeding/snack as they should not go 10-12 hours without eating. If the FOD child/adult has experienced hypoglycemia (even with frequent meals), in order to help decrease the frequency of low blood sugar in the morning, some find it helpful to mix in 1-3 tablespoons of raw cornstarch (i.e., Argo) to a cold sugar-free liquid or food at night. Be aware that too much cornstarch can be constipating for some and may be hard on teeth. Be sure to brush after eating or drinking it. Also note that cornstarch should not be given to an infant under age one (they lack the necessary pancreatic amylase to digest the cornstarch).

Some physicians also may prescribe an L-carnitine supplement (FDA-approved Carnitor® or an FDA-approved generic form—these are NOT the same as the healthfood store carnitine), especially if a secondary carnitine deficiency is involved. Although carnitine is made within the body, sometimes supplementation is necessary. It is useful in stabilizing blood sugar levels, increasing muscle tone, and removing metabolic waste products from the cells so a buildup of toxins will not occur.

As with any medical condition, discussing treatment options with your physician(s) and FOD specialists is necessary for the well-being of anyone with an FOD. Please do NOT make any changes in your child's or your own treatment without first consulting with your physicians.

Early Screening, Diagnosis and Treatment will prevent needless deaths

Disorders of Fatty acid Metabolism

Medium Chain acyl coA Dehydrogenase Deficiency (MCAD), Medium Chain 3-Ketoacyl coA Thiolase Deficiency (MCKAT), Medium/Short Chain L-3 Hydroxy acyl coA Dehydrogenase Deficiency (M/SCHAD), Long Chain L-3 Hydroxy acyl coA Dehydrogenase Deficiency (LCHAD), Very Long Chain acyl coA Dehydrogenase Deficiency (VLCAD), Short Chain acyl coA Dehydrogenase Deficiency (SCAD), 3-Hydroxy acyl coA Dehydrogenase Deficiency (HADH, formerly called SCHAD), Short Chain 3-Ketoacyl coA Thiolase Deficiency (SKAT, 3-ketothiolase), Electron Transfer Flavoprotein and Electron Transfer Flavoprotein ubiquinone-oxidoreductase Deficiency (ETF and ETF-QO, also known as GAII or MADD), Carnitine Palmitoyltransferase Deficiency (CPT I & II), Trifunctional Protein Deficiency (TFP), Carnitine Acylcarnitine Translocase Deficiency (CACT), Carnitine Transport Defect (Primary Carnitine Deficiency), 2,4-Dienoyl coA Reductase Deficiency, 3-Hydroxy 3-Methylglutaryl Deficiency (HMG), Unclassified FODs

Additional Metabolic Family Support Resources

Organic Acidemia Association (OAA)

Kathy Stagni, Director
13210 35th Avenue North
Plymouth, MN 55441
(763) 559-1797
(866) 539-4060 fax
http://www.oaanews.org
oaanews@aol.com

United Mitochondrial Disease Foundation (UMDF)

8085 Saltzburg Road, Suite 201
Pittsburgh, PA 15239
(412) 793-8077
(412) 793-6477 fax
http://umdf.org
info@umdf.org

Institute of Metabolic Disease Baylor University Medical Center

Supplemental NBS & FOD Diagnostic Testing

Charles Roe, MD, Medical Director
3812 Elm Street
Dallas, TX 75226
(214) 820-4533 (Diagnostic info)
1–800-4-BAYLOR (NBS info)
(214) 820-4853 fax
http://www.baylorhealth.edu/imd/Default.htm

Children Living with Inherited Metabolic Disease (CLIMB)

Climb Building
176 Nantwich Road
Crewe CW2 6BG
United Kingdom
0800 652 3181
0870 7700 327 fax
http://www.climb.org.uk
Info.svcs@climb.org.uk

Children's Mitochondrial Disease Network

EMDN, Mayfield House
30 Heber Walk, Chester Way
Northwich CW9 5JB
United Kingdom
440 01606 43946
http://www.emdn-mitonet.co.uk
Info_cmdn@btopenworld.com

Sigma-Tau Pharmaceuticals, Inc. Makers of Carnitor®

800 South Frederick Avenue, Suite 300
Gaithersburg, MD 20877
(800) 447-0169
http://www.sigmataupharma.com
sigmatauinfo@sigmatau.com

You are not alone...