

## Practical Nutritional Considerations in Organic Acidemias

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### Introduction

Decisions regarding the medical and nutritional care plan for a child or adult diagnosed with a disorder of organic acid metabolism (Glutaric acidemia type I (GA I), Isovaleric acidemia (IVA), Methylmalonic acidemia (MMA), Propionic acidemia (PPA), and 3-Methylcrotonyl-CoA carboxylase deficiency (3-MCC def'y) is a team effort. Parents and caregivers are often expected to take on the role of team leaders in creating a 'Medical Home'<sup>1</sup>. Input from the patient, parents, and caregivers is required most of all, in order to meet the daily medical and nutritional needs of OA patients. Therefore, ongoing communication with your primary care physician, metabolic physician, and metabolic clinic staff is key. Keeping the lines of communication open with your metabolic clinic will best assist them to work with you to tailor the OA diet to your child's needs and adjust the diet as medically necessary to optimize metabolic control, growth and development, and maintain and support good health. If there are multiple daily caregivers (parents, relatives, teachers, nurses, sitters, etc) it is important for everyone to be consistent and aware of the most current OA nutritional plan, in order to avoid errors or miscommunication.

Patients, parents, and caregivers of individuals living with an OA and other metabolic disorders deserve great respect and admiration for their dedication and daily efforts. As we all know, coordinating the medical nutrition therapy for an individual diagnosed with a metabolic disorder such as an OA is a challenge. In planning the metabolic nutrition diet orders for an individual patient, take into consideration individual, age-specific nutritional needs, as well as the practical issues that might influence or hinder the actual achievement of the diet goals. Consider each individual's short and long term dietary requirements and goals, and target metabolic nutrition education counseling to the needs and abilities of each individual living with an OA. It is important to strive for realistic and achievable goals. There can be big differences in the nutritional tolerance and needs of individual OA patients.

OA patients have distinct nutrition and feeding issues and needs at different times and stages in their lives. Some OA patients do well meeting their daily nutritional needs via oral feedings (PO), while others rely on tube feedings (TF) or a combination of PO and TF routes, when in good health. During times of illness, most OA

when in good health. During times of illness, most OA patients often require the support of TF and/or intravenous (IV) fluid therapy to assist in illness recovery and regain metabolic control. OA dietary goals and restrictions can be very different between individual OA patients, and for each OA patient in different medical situations at different times of his or her life. An individual's OA diet plan depends upon his or her diet tolerance; medical and nutritional needs for growth, development, and/or health maintenance; biochemical lab results; and medical/illness status. I would urge you to continue to discuss your individual questions and concerns with your metabolic clinic physician, dietitian and staff.

### Overview

There are certain practical considerations that must be addressed in order to optimize an OA medical nutrition therapy plan to ensure the best possible outcome (Table 1).

The ongoing availability of these medical services and

#### ***OA Nutrition Practical Considerations: Availability of.....Table 1***

***State Department of Health Newborn Screening  
Program for OAs***

***Metabolic Physician Services and Staff***

***Health Insurance Coverage/State Legislation***

***State Department of Health Assistance Programs***

***OA Medical Foods (formula) & Low protein Specialty Foods***

***Supplies: Digital Gram Scales, Books (amino acid/protein food counts, cookbooks)***

***L-amino acid, vitamin supplements (l-valine, l-isoleucine, hydroxycobalamin, etc)***

***Local Outpatient Lab Draws and Home Monitoring (urine ketones)***

***Emergency Illness Management Protocols***

***Support for OA Diet at School (and/or Work)***

nutritional supplies are important tools that will assist you in implementing the OA diet on a daily basis. Patients who do not have continuing access or limited coverage of these items often have difficulty adhering to

OA diet goals.

### **Calling your Health Insurance Company**

Medical health insurance coverage of OA Medical Foods (formulas) and as needed, Low protein specialty foods are often key issues. If you haven't yet had difficulties with your health insurance regarding coverage of formula and/or low protein specialty foods, unfortunately chances are high that you may encounter some problems in the

<b><i>Health Insurance Coverage Issues: Denials....Table 2</i></b>
<b><i>Find out WHY? Request it in writing.</i></b>
<b><i>Find out how to Appeal: How long it takes, how/who will get response</i></b>
<b><i>Request your metabolic clinic to provide Letter(s) of Medical Necessity</i></b>
<b><i>Are you eligible for 2<sup>nd</sup>ary state Medicaid insurance coverage</i></b>
<b><i>Are you eligible for your state Department of Health assistance programs</i></b>
<b><i>Contact your state Department of Insurance to check if your health insurance is in compliance with state laws (if available)</i></b>
<b><i>Ask your benefits office if your insurance is a 'self funded plan' exempting them from following state insurance mandates (ERISA) even if you have a state law</i></b>
<b><i>Contact your local and state legislators for assistance</i></b>
<b><i>Consider obtaining assistance from a legal advocate</i></b>
<b><i>Consider changing your health insurance provider if possible</i></b>
<b><i>Contact your metabolic clinic staff to avoid running out of supplies</i></b>

future (Table 2).

If your health insurance offers case management with an assigned caseworker (usually a nurse) or if you can find a helpful member services contact person to assist you with your insurance coverage issues, this can save you some time and effort in the future. It is very important to understand your health insurance coverage plan benefits, policies, and limitations, and stay informed of any policy changes over time.

Your need to be diligent is not ended even when you get approval for health insurance coverage. Some health insurance providers require periodic reassessments and regular prior authorizations for coverage of OA Medical Foods, Low protein specialty foods, and uncommon medications. Ask if your authorization has an expiration date in order to be prepared to renew it in the future, and keep track in your calendar to allow for enough time (4 weeks or more) for the renewal process. Ask if your OA Medical Foods are covered under your medical benefits (durable medical equipment or home health infusion) or prescription benefits. Your policy coverage may vary, have limitations, or have different co-pays or deductibles;

depending upon the type of coverage that your OA Medical foods, low protein specialty foods, L-amino acids, and vitamin supplements are placed under by your health insurance company.

Who will supply your OA Medical Foods, Low protein specialty foods, and OA specific medications?

### **OA Medical Foods**

If covered under your prescription policy, you may find it best to order your OA Medical Foods from a local pharmacy and make the monthly trips to pick it up. This is convenient, as long as the pharmacy can do direct insurance billing; sparing you the need to pay out of pocket, submit insurance claims with your receipt, and wait for reimbursement. Depending upon your insurance policy and state laws (if applicable) you may have out of pocket co-pays or deductibles for your OA Medical Food prescriptions.

If covered under your medical policy, some families fill their OA Medical Food prescriptions via home health infusion (HHI) or durable medical equipment (DME) companies contracted with their health insurance, and have delivery services to their home. This option depends upon your medical insurance benefits and coverage. Insurance companies often contract with different home health companies, therefore you may want to ask your insurance for a list of approved HHI or DME providers. Usually your metabolic clinic staff must first contact the HHI/DME to provide prescriptions, refill authorizations, medical necessity letters, and set up the initial deliveries to your home. Often individual HHI or DME companies have limitations in terms of what they can provide. Most contracted HHI or DME companies can order the metabolic medical foods and bill your insurance. Some companies are limited by company policies or state license to provide only 'enteral feeding supplies', meaning supplies necessary to support a patient getting their metabolic medical food by nasogastric (NG), gastric (G), or jejunum (J) tube feedings. Home infusion companies are DMEs with state licensed pharmacies, and are able to assist patients who require IV medications or nutrition support. Some HHI or DME companies can not assist patients who are drinking all of their formula by mouth, because of their company policy limitations. Some HHI or DME companies require caregivers to place a monthly call to order refills, while others will automatically send out monthly refills. Your metabolic clinic staff and health insurance case coordinator should be able to help you sort out the best option for your family.

### **Low protein specialty foods**

In terms of the Low protein specialty foods, some state laws specify limitations of coverage per year. For exam-

\$2500 per year per metabolic patient for low protein specialty food health insurance coverage. Most families direct order and pay the low protein food companies, then submit claims to their health insurance and wait for reimbursement. It is very difficult to find a Pharmacy, HHI, or DME company willing to order and successfully bill health insurance companies directly for the low protein specialty foods. In some cases, depending upon their insurance policy, families are able to ask the low protein food companies to submit the bill directly to your health insurance company. But not all low protein food companies provide this service. This is a very convenient service if available, but typically requires many calls and a lot of your time spent filling out forms, however caregivers often find it worth the effort.

**OA specific medications**

Some pharmacies are compounding pharmacies, where the pharmacists are able to order manufactured medications (L-amino acids, vitamins) in tablet or powdered form and prepare them with water or add flavorings to increase taste acceptance for PO or TF administered medications. The pharmacist may be able to mix several medications into one liquid medication as chemically allowable. It is important to know your pharmacy insurance benefits, copays, and limitations. Ask your insurance for a list of contracted compounding pharmacies in your area. Your metabolic physician will need to send the prescriptions.

Finally, a few states still have department of health sponsored programs to assist families with obtaining metabolic formulas, low protein specialty foods, and other metabolic medical supplies; e.g. Massachusetts PKU and Other Metabolic Disorders Special Dietary program, Maine’s Children with Special Health Needs program, Illinois Division of Specialized Care for Children program, your state’s Women, Infants and Children (WIC) program, or your state Medicaid program. But most programs have eligibility requirements - state residency, diagnosis, financial need, written insurance denials for coverage, and/or patient’s age. Discuss with your metabolic clinic staff to find out if you are eligible for a state program that may assist you.

**What to do if your insurance denies coverage?**

Should you get insurance denials for coverage (Table 3), research the appeals process and policies from your insurance company. You can request assistance from your metabolic clinic staff with phone calls and letters of medical necessity for more in depth insurance medical review. This can be a frustrating process that may take some time for review and decisions. It is important to continue to be as patient and courteous as you can in your interactions with your insurance company.

**Health Insurance Coverage: When Calling your insurance...Table 3**

*Try to be patient and courteous, inform and educate, try to gain an advocate*

*Review your Health Insurance plan coverage benefits and limits*

*Request case management services if available to assist in care coordination and problems*

*Document each call and contact person, info, department, phone numbers (extension), date of call*

*Request information in writing for future reference*

*Request a copy of your Explanation of Benefits (EOB)*

*Refer to OA formulas as ‘OA Medical Food’, prescribed by your metabolic MD’\**

*Know your State Law<sup>2</sup> and it’s limits (if available) and have a copy in your files*

*\*Exception: Always refer to OA Medical Foods in the same terms as your state law as applicable*

Try to consider your correspondence with your insurance company as an opportunity to educate and inform insurance workers about OA and the strict and difficult medical nutrition therapy requirements for patients. By outlining what you and your family have to go through, you might gain an ally at your insurance company who is willing to assist you in finding a solution to these (and other) problems and concerns. Whenever possible, avoid taking a belligerent approach, instead try to be polite but persistent. If you still have questions and the health insurance staff person seems unsure or unable to provide answers, request to speak to a supervisor to request further clarification.

Appendix I contains a sample medical necessity letter that may assist you in your insurance appeals process. You might need to seek the assistance of a legal advocate to support your appeals if you continue to get illegal denials for coverage in spite of all of your best efforts. If your employer offers a ‘self funded’ insurance plan, the ERISA (Employee Retirement Income Security Act) exemption may be an obstacle to getting coverage; this apparently exempts the employer/health insurance from having to follow certain state insurance mandates. You must then appeal directly to your employer for coverage (through your benefits office) on the basis of medical necessity for assistance. Mention your state law if available and the American Academy of Pediatrics’ 1994 position statement<sup>3</sup> encouraging health insurance coverage as support for coverage. You may also request an independent medical review in the appeals and review process, ask your Department of Insurance Consumer contact for assistance. A few families have had success with the support of local or state legislators in reversing

insurance coverage denials, even without a state law. But if your state doesn't currently have a state law mandating health insurance coverage for medical foods or low protein specialty foods; then contacting your local state and national parent support groups to assist in organizing grassroots efforts in your state to get a law passed by your state legislators has been successful in several states. Providing your legislators inarguable information on medical necessity and financial burden of medical costs without insurance assistance will often win political support and sponsorship of a bill. If all else fails, it may be necessary to look for a different insurance provider.

If changing your insurance carrier is a consideration, make sure to do your homework to check the benefits of the new plan. Watch out for pre-existing condition exemptions, and ask how they would provide for your OA medical needs before finalizing the change. The federal law, Health Insurance Portability and Accountability Act (HIPAA) of 1996 (sometimes called "Kennedy Kassebaum") might be helpful to continue to obtain coverage and avoid pre-existing conditions clauses (Reference 1). It is also important to know if your health insurance plan sets age limits (eg. 18 yrs old) for coverage of dependent children. "Given the wide variation [among state laws and individual policies], it is recommended to contact your insurance company or plan sponsor at least several months before the child turns age 19 to tell them that they have a dependent adult child with a disability....[and it is critical to obtain state-specific advice]." <sup>4</sup> Without updating your insurance company, your child's coverage will automatically stop at the age specified by your plan. Young adult patients who lose insurance coverage because of age limitations, can ask their state department of insurance for information on applying for individual coverage as provided by this HIPAA federal law. If you lose your health insurance coverage, you should also discuss other options with your social worker or financial coordinator at your primary care physician's office or metabolic clinic. The cost of medical care for treatment of OA is financially overwhelming for most caregivers and patients without health insurance coverage or with limited coverage.

### **Getting Organized**

It takes much of your time and effort to have all of your various medical services organized, insurance coverage verified, and in place. I would recommend a methodical and step-wise approach to finding solutions to challenges that you may face in order to ensure that your child obtain the medical and nutritional support and services needed to promote good OA metabolic control.

Parents and caregivers of individuals with special health care needs are now expected to take on increased responsibility for coordinating their child's care. Caregivers often must work with multiple institutions and agencies; pri-

mary care physicians and staff, specialty physicians and staff, hospitals, outpatient labs, state department of health programs and case workers, health insurance workers, home health supply companies, durable medical supply companies, pharmacists and staff, visiting nurses, etc. It can be overwhelming for many caregivers and patients to organize all of the contacts and information. Therefore having an OA Individual Health Plan (IHP)<sup>5</sup> (Appendix II), along with a written OA emergency illness protocol may help both caregivers and medical and specialty physicians and staff in organizing and recording important contacts and medical/nutrition issues. However, it requires regular updating.

### **Summary**

Staying in touch with your metabolic clinic physician, dietitian, and staff should best assist you with adjusting OA medical nutrition therapy goals for your child and his or her changing needs over time. Obtaining and maintaining health insurance coverage of OA Medical Foods, Low protein specialty foods, and L-amino acid medication supplements can be very complicated. It is important to know your health insurance policy benefits and limitations; what your state law allows, and other state program resources. Ask your primary care physician and your metabolic clinic physician and staff for information to help you decide which resources are most helpful for your situation. Coordination of your child's OA medical and nutritional care can be one of your biggest challenges; organizing your information and contacts can greatly assist you and save your time. I would like to thank Dr. Mark Korson for reviewing this article, and most of all, thanks to the parents and individuals living with OA whom I have been privileged to work with and from whom I have learned the most.

### **Reference:**

1. Website: Medical Home Initiatives for Children with Special Health Care Needs. American Academy of Pediatrics. [www.aap.org](http://www.aap.org) (Community Pediatrics).
2. Website: National PKU News: State Laws and Policies by Virginia Schuett. [www.pkunews.org](http://www.pkunews.org) (PKU Legislation and Policies) or contact your state government.
3. *Pediatrics*, Vol 93 (5): May 1994, p. 860
4. Schulzinger, R. 2000. *Youth with Disabilities in Transition: Health Insurance Options and Obstacles*. An occasional policy brief of the Institute for Child Health Policy, Gainesville, FL (ISBN: 0-9700909-4-3) or ([www.mchbhrw.org/materials](http://www.mchbhrw.org/materials))
5. Silva TJ, Sofis LA, and Palfrey JS. 2000. *Practicing Comprehensive Care: A physician's operations manual for Implementing a Medical Home for Children with Special Health Care Needs*. Boston, MA: Institute for Community inclusion/UAP, Boston. ([www.childrenshospital.org/ici](http://www.childrenshospital.org/ici))

**Appendix I: Health Insurance Sample Letter of Medical Necessity for PPA\***

\* **Change all underlined sections to fit your individual case.**

January 3, 2003

Attn: RN Case Coordinator or MD Medical Director Name  
Medical Appeals Review  
Health Insurance Company Name  
Phone # and Fax #

Re: A.B. DOB: 12/20/92 (replace with patient's name, date of birth, insurance #)  
Insurance ID #123456789

Dear Case Coordinator;

*The purpose of this letter is to document the medical necessity for A.B. to receive his metabolic enteral formula ingredients; PPA Medical Foods, L-amino acid solutions (10mg/ml), carnitor, as well as his Gastrostomy tube feeding and medication administration supplies; Enteral feeding pump, Enteral feeding bags, MIC-KEY G button, G-tube feeding extension sets, Luer lock syringes, tape, and urine ketone testing strips.*

*A.B. has been diagnosed with a metabolic disorder called Propionic Acidemia (PPA). Medical treatment of PPA patients requires careful medical monitoring with a strict dietary restriction of the amino acids; L-valine, L-isoleucine and L-threonine and L-methionine, because of the metabolic impairment of Propionyl-CoA carboxylase enzyme. A.B.'s daily dietary intake of these four essential amino acids are restricted to the minimum amount he needs to promote continued growth and development, while also preventing PPA related metabolic imbalances. It is also very important to try to avoid catabolism and imbalances by ensuring daily intake of adequate total calories and providing enough of the other essential amino acids. Metabolic instability for PPA patients could result in acute metabolic decompensation, delayed growth, delayed development, neurologic impairment, and potentially require increased frequency of inpatient admissions for medical management and in prolonged cases can lead to death.*

*We prescribe PPA Medical Foods, L-amino acid solutions, in order to provide A.B. with minimal amounts of the amino acids; L-valine, L-isoleucine and L-threonine, L-methionine; while still providing adequate amounts of the other essential amino acids, as well as carbohydrate, essential fatty acids, vitamins and minerals. We routinely adjust A.B.'s prescriptions based upon his biochemical lab results, clinical status, growth and developmental needs, as well as with his daily nutritional requirements for his age in mind.*

*A.B. receives 90-100% of his nutrition via his Gastrostomy tube feeds on a daily basis. Therefore, he requires a daily supply of enteral feeding bags, a reliable enteral feeding pump, and other G-tube supplies as listed above. His daily liquid medications, L-amino acids and carnitor, are all administered to him by measured doses in syringes via his G-tube. The urinary ketone sticks are used by A.B.'s caregivers at home to help assess his metabolic stability during the day and especially during an acute illness, this information assists us in making home metabolic therapy decisions.*

*We request your approval for coverage of his prescriptions for; PPA Medical Foods, L-amino acid solutions (10mg/ml), carnitine (100 mg/ml), as well as his Gastrostomy tube feeding and medication administration supplies; Enteral feeding pump, Enteral feeding bags, MIC-KEY G button, G-tube feeding extension sets, 60 cc and 12 cc Luer lock syringes, tape, and urine ketone testing strips. These supplies are essential to A.B.'s continued PPA metabolic stability. As you know, coverage of medically necessary enteral formulas and food products modified to be low protein for individuals with inherited diseases of amino acids and organic acids are mandated in the Commonwealth of Massachusetts by the passage of the Law of 1994 chapter 384 (House Bill #5622). The American Academy of Pediatrics in the 1994 position statement recommends that these 'special medical foods used in the treatment of amino acid...disorders are medical expenses that should be reimbursed.'<sup>1</sup> With appropriate treatment and follow up, we expect that A.B. will continue to stay metabolically stable and achieve his growth and development potential within the limits of his PPA. Because of the nature of his disorder and medical concerns, and in order to insure the purity and accuracy of his L-amino acid dose, we require that a licensed compounding Pharmacist prepare and dispense this specially prepared amino acid solutions to A.B.*

If you have any questions regarding any of this information, please contact us at (123) 456-1234. We would appreciate it if you could inform us of your decision on this matter as soon as possible. A.B. will need refills of his medical supplies by the end of this month. With his previous insurance policy, he was obtaining his medications from the ABC Pharmacy and XYZ Home health supplies. Please inform us of the how best to proceed with his new insurance policy requirements. Thank you for your time and your assistance in coordinating the care of this patient.

Sincerely,

Metabolic Physician, M.D.

Metabolic Dietitian, R.D.

Division of Metabolism

Division of Metabolism

cc: A.B.'s caregivers

<sup>1</sup> Pediatrics Vol. 93 No.5 May 1994 p86