

Diets for Inborn Errors of Metabolism

Keeping Track

More than counting calories.

Most metabolic conditions require very specific diet restrictions. In some cases the diets are quite limited. Finding tasty and nutritious food that follows the diet can be challenging. In addition, the diets often require carefully measuring portion sizes of certain types of food to make sure the amount stays within safe levels. Over time, these tasks can become second nature, but initially it takes a lot of work.

Diet Records

Keeping a daily journal helps you stay on track. Even after the diet becomes routine, it is still necessary to keep written diet records for health care providers to review. These written records help your provider monitor eating patterns over time. Diet records also give important information when combined with blood and/or urine tests. So before having certain blood or urine tests, you may be asked to complete a [3-day diet record](#).

How to Measure Foods

Foods can be measured using either standard or gram measurements. Measuring in grams requires the use of a special gram scale.

Using a Gram Scale

A gram scale is the most accurate way to measure foods and metabolic formulas. This is very important for people on protein restricted diets. It is also better for weighing foods that are difficult to measure with measuring cups and spoons, such as cereals and other lumpy or chunky foods. You can find gram scales at most kitchen stores or order one online.

Using Standard Measurements

If a gram scale is not available, standard measurements can be made using typical kitchen utensils. These include: standard measuring spoons, standard measuring cups, and a standard glass measuring cup. Do not use non-standard utensils such as coffee cups, drinking glasses or serving spoons. All measurements must be lightly packed and level.

This table has comparisons between common standard measurements that can be helpful when calculating portion sizes

Tablespoons (T)	Cups (c)	Teaspoons (t)	Fluid ounces (fl oz)	Quarts (qt)
1 T		3 t	½ fl oz	
2 T	1/8 c		1 fl oz	
4 T	1/4 c		2 fl oz	
5 T + 1 t	1/3 c			
8 T	1/2 c		4 fl oz	
12 T	3/4 c		6 fl oz	
16 T	1 c		8 fl oz	1/4 qt
	2 c		16 fl oz	1/2 qt
	4 c		32 fl oz	

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Metabolic Formulas

Special formulas for good health.

People with metabolic disorders usually have restricted diets. This can make it hard to get all the nutrients you need. When nutrients are missing in the diet, they must be added in another way. We do this by using special formulas or supplements or both.

How to Get Metabolic Formulas

Metabolic formulas can have very different amounts of protein, calories, vitamins and minerals. Your dietitian will work with you to find the best one to treat your condition and meet your health needs. Once we know which formula you need, a Regional Metabolic coordinator places the order and has it delivered to your home. To order more formula, just call a Program coordinator. Re-orders should be placed while you still have about a 2-week supply. It is important to call BEFORE you run out of formula because re-orders can take up to 2 weeks to arrive.

SPECIAL NOTE: If you have problems getting your order, contact a Program coordinator by phone or email. Be aware that phone and email messages are only checked Monday through Friday during business hours.

Phone: (510) 752-7703

Email: metabolic-program@kp.org

Making Metabolic Formulas Taste Better

It can be tough to drink the same formula every day. Metabolic formulas can be mixed with different ingredients to make them taste better and add some variety. Examples of things to add: pureed or frozen fruit, nondairy creamer, flavored extracts, Mocha Mix, Kool-Aid, or Tang. If you still have trouble taking the formula as prescribed, do not give up. Talk with your dietitian about possible solutions.

To order more metabolic formula
CALL
(510) 752-7703

Diets for Inborn Errors of Metabolism

Ordering Metabolic Medications

Medications come to you.

We have a special Metabolic Pharmacy at Kaiser Permanente to make it easy to order and receive your metabolic medications. Metabolic medications must always be ordered through the Metabolic Pharmacy.

This is how it works:

A program coordinator places the Initial order for metabolic medications with the Metabolic Pharmacist. Once the order is set-up you can get refills as needed.

For refills, call the Metabolic Pharmacy Refill Line: (415) 833-2698.

A recording will ask for the following information:

Patient Name

Medical Record #

Name of Medication

Prescription #

Phone number where you can be reached (9 am - 5 pm that same day).

Any specific instructions regarding delivery.

There is no need to wait in line for the medication. Orders are mailed directly to you.

For orders that have a co-pay, your credit card is charged upon delivery.

IMPORTANT NOTE: ALWAYS keep a 2-week supply of your medication(s) on hand. It sometimes takes up to 10 days to get refills. If you have any questions or problems with this system, please contact the Regional Metabolic Clinic at (510) 752-7703.

For refills on metabolic medications
CALL
(415) 833-2698

This page applies ONLY to those who have a metabolic disorder that requires a low protein diet.

Disorders requiring low-protein diets fall into three categories:

Amino Acidemias

Examples: PKU, MSUD, homocystinuria

Organic Acidemias

Examples: MMA, PPA

Urea Cycle Disorders

Examples: OTC Deficiency, Lysinuric Protein Intolerance

Diets for Inborn Errors of Metabolism

Low Protein Specialty Foods

Variety is the spice of life.

Following a low-protein diet can be hard. One way to make it easier is to find new foods that still follow the diet. For example, rather than avoiding all breads and pastas, try using low-protein breads and pastas. You can also find websites with low-protein recipes.

Low-protein specialty foods help add variety to your diet. Kaiser Permanente has contracted with several specialty food companies. These companies offer specialty foods for patients that require a low-protein diet. These specialty foods can be ordered by patients who are seen in the Regional Metabolic Program.

To order call the Regional Metabolic Program coordinator (510-752-7703) and ask for the order forms. Fill out the forms and return them to the clinic. The Program coordinators will review the form and place your order. The order is delivered directly to your home.

SPECIAL NOTE: All orders must be placed by the Regional Metabolic Program. Kaiser will not pay for orders placed directly with the vendors.

Contracted Food Companies

Kaiser has contracts for low-protein foods with the following food companies. You can visit their websites to see what is available. Check back often because products change frequently:

[Cambrooke Foods](#)

[Dietary Specialties](#)

[Ener-G Foods](#)

[Nutrica/SHS North America](#)

[Maddy's](#) (Applied Nutrition)

[PKU Perspectives](#)