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Medical Nutrition Therapy for Pediatric Kidney Stone Prevention, Part 3: Cystinuria

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Audience: Pediatrics; Nephrology Outpatient Clinic

CYSTINURIA IS AN inherited, genetic condition that causes hyper-excretion of cystine and other dibasic amino acids (lysine, arginine, and ornithine) in the urine due to impaired transport in the proximal renal tubules. The low solubility of cystine leads to precipitation and the formation of renal stones.[1,2] While rare, this condition accounts for a relatively significant proportion of pediatric nephrolithiasis, up to 10% in children compared to 1% in adults.[3,4] Furthermore, as compared to other causes of pediatric stone formation, cystinuria is more likely to result in frequent recurrence, morbidity, and need for medical interventions. Careful attention to disease management is required to prevent disease-related complications, specifically chronic kidney disease.[1,4]

The dietary management of cystinuria is a two-pronged approach involving (1) a reduction in the overall urinary excretion of cystine and (2) an increase in urinary cystine solubility. The first objective may be obtained by restricting dietary intake of animal proteins, which, in comparison to plant-based proteins, are generally higher in cystine and its metabolic precursor methionine.[2,3] Avoiding excessive dietary intake of sodium will also reduce urinary excretion of cystine.[3,4] The second objective, increased urinary cystine solubility, can be improved with increased fluid intake; in pediatric patients, this is commonly estimated using body surface area and approximates 2 L/m^2 × body surface area.[3,5] Cystine solubility also increases with urinary alkalinity, so once again it is important for patients to avoid excessive animal protein, whose sulfur containing amino acid profile acidifies the urine.[3,6]

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Finally, it is noted that patients with cystinuria are at higher risk for calcium oxalate stones; therefore, general dietary recommendations for renal stones should be followed, including ensuring adequate amounts of dietary calcium.[2]

Positive outcomes have been noted with an increase in vegetable-based protein and limitation of animal based to 60% of dietary protein intake, including at least two sources of dairy foods.[2] While randomized clinical trials are the gold standard to confirm such interventions, clinical studies are rare for nutritional therapies and even more so for pediatric populations. The overall health benefits and proven safety of balanced plant-based diets lend itself toward implementation with available evidence—albeit with special precautions to ensure the optimal growth and development of children. As for the safety and efficacy of such diets in children, it is the position of the Academy of Nutrition and Dietetics that vegetarian diets, when well planned, "are appropriate, and they satisfy the nutrient needs and promote normal growth at all stages of the life cycle, including pregnancy and lactation, infancy, childhood, [and] adolescence…"[7] That said, it is important to note that removing dietary protein, without substitution with appropriate vegetable protein, can, in fact, be harmful to the growth and development of pediatric patients. A registered dietitian should consult with the family to assess the child's usual intake and nutrition status, as well as food access and social or economic barriers to making dietary changes. Modifications should be appropriate to the patient and caregiver's mealtime routines and level of culinary expertise. A simple recommendation might be to suggest a single food swap that is easy and affordable; examples may include oatmeal or yogurt in place of bacon at breakfast, a peanut butter sandwich substituting for ham at lunch, or bean instead of beef burritos for dinner. Table 1 provides protein intake recommendations for pediatric patients, allowing for a range depending on food choices. Higher protein recommendations are recommended for children limiting animal products (e.g., vegan diets). Children who consume lacto-ovo-vegetarian diets generally meet or exceed recommended protein intake, but clinical judgment as to the nutrition status of the patient is advised.

Table 1. Recommended Daily Protein Intake Based on Age

Age Group	Lower Protein Requirement (g/kg/d)	Higher Protein Requirement (g/kg/d)*
Ages 1–3 years**	1.1	1.5
Ages 4–8	0.95	1.24
Ages 9–13	0.95	1.15
Ages 14–18	0.85	1.02

*Higher protein recommendations for children limiting animal products (e.g., vegan diets).

**Recommendations are estimates for children above 13 months of age.[7-9]

Finally, certain vitamins and minerals, and specifically B-12, should be supplemented in any child who regularly avoids or limits animal products.[7]

In conclusion, protein should not be restricted below the dietary reference intakes in growing children; however, protein foods lower in methionine, such as plant-based foods, may be encouraged. High animal protein diets should be avoided. A registered dietitian can help parents to select low-methionine protein foods and provide recommendations for vitamin supplementation if needed. The educational handout that follows provides a sample list of common foods categorized by methionine content. The following handout can be used to emphasize the differences in methionine content within and among food groups, but should not be used as a recommended food list. For example, all protein foods are presented in two ounce equivalents in order for appropriate comparisons to be made regarding methionine content per serving.

Note to Educators

All handouts provided in this series should be utilized as tools to facilitate communication between providers and caregivers and should in no way substitute for complete and thorough medical care.

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Diet Tips for Cystinuria

If your child is suffering from cystinuria, this handout will help you to select foods lower in methionine. The numbers below are estimates of methionine content in common foods.
These are examples only. This list does not substitute for medical advice.

Estimates of Methionine Content in Common Foods

Low Methionine: <100 mg		**Medium Methionine: 100-300 mg**
Apple: 2	Grapes (1 c): 19	Refried beans: 100
Berries: 2-10	Broccoli: 34	Cashews: 105
Cucumber: 3	Sweet potato: 42	Black beans: 110
Lettuce: 3	Almonds: 45	Soybeans (edamame): 110
Pear: 4	Corn: 50	Tofu: 135
Watermelon: 4	Peas: 60	Sunflower seeds (2 tablespoons): 140
Tomato: 5	Potato: 60	Surimi (imitation crab): 150
Banana: 9	Baked beans: 60	Cheese, cheddar (1 oz) 155
Macadamia nuts: 10	Soymilk (1 c): 65	Cheese, mozzarella stick: 195
Kale, cooked: 11	Walnuts: 70	Vegetarian burger: 200
Carrots: 13	Oatmeal (1 serving): 71	Milk, 1% (1 c): 215
Green beans: 15	Lentils: 75	Yogurt, low fat, fruit (6 oz): 245
Mushrooms: 17	Peanut butter (2 tablespoons): 85	Shrimp (8 large): 295
High Methionine: >300 mg		
Brazil nuts: 315		Beef, lean, ground patty: 475
Canadian bacon (2 slices): 316		Chicken breast: 490
Fish, salmon: 335		Lobster, tail: 530
Eggs (2): 390		Crab, cooked: 730
Fish, canned tuna: 445		Pork chop (1 chop): 1500
Turkey, roasted: 450		Pork, cured ham, (1 slice): 2230

Amounts listed above are estimates of methionine content. Unless otherwise stated, foods are listed in standardized portion sizes: ½ cup serving or medium size whole fruit or vegetable,1 oz nuts, 2 tablespoons peanut butter, 2 oz meat, ½ cup beans. (SOURCE: USDA Nutrient Database Release 28)

- Limit animal-protein to one serving per day. These foods are generally high in methionine.
- Encourage protein from plant-foods, such as soy, beans, and nuts.
- Include two servings of dairy or calcium-rich foods in your child's diet each day.
- Processed meats such as bacon, sausage, pepperoni, and pre-packaged deli meats are high in both methionine and sodium, which can worsen cystinuria.
- Your child needs protein to grow and develop. Grams of protein are listed on food labels (pointed out on the sample to the right). Talk to your dietitian about your child's daily protein needs.
- Discuss any changes to your child's diet with his or her physician.

Your child needs _____g of protein each day.