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During fasting, the body initially uses mostly stored glycogen (starch) from the liver to meet energy needs. After the glycogen stored in the liver is used up, muscle proteins then become the main source of energy. A longer fast will mean that more muscle protein will be needed to provide energy.

In Barth syndrome, people tend to use a larger percentage of their muscle mass during an overnight fast. This can make it difficult to maintain or gain muscle mass.

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**Liver Glycogen**

(Starch)

Main source of energy **during**  
the first six hours of fasting

—————▶ Energy

**Muscle Protein**

Main source of energy **after** six  
hours of fasting.

—————▶ Glucose

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Uncooked cornstarch is the most slowly absorbed carbohydrate. It takes between 4-6 hours for the cornstarch to be completely digested. Because the cornstarch is slowly digested, the amount of time spent fasting is decreased after eating cornstarch. This helps to prevent hypoglycemia and minimize muscle protein losses overnight.

**When should I use Cornstarch?**

- Cornstarch should be given before bed.
- Infants can be given cornstarch once they are sleeping more than 8-9 hours on a regular basis.

**How Much Cornstarch is Needed?**

The goal is to provide the amount of calories needed during the first 4-6 hours of sleeping. The amount of calories needed is based on the weight of the child. The following table provides a general guideline for dosing.

**Cornstarch Fact Sheet**

The Barth Syndrome Foundation [www.barthsyndrome.org](http://www.barthsyndrome.org)  
Rhonda Avery RD, LD/N,CDE (Watson Clinic LLP) June 2006

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Cornstarch Dosing			
Age	Weight kg	Weight lbs	Cornstarch
12 – 18 Months	10	22	2 teaspoons
18 – 36 Months	12.5	27.5	3 teaspoons
3-5 Years	15	33	3.5 teaspoon
5 – 8 Years	17	37.5	4 teaspoons
8 – 12 Years	22	48.5	5 teaspoons
12 – 15 years	30	66	2 tablespoons
Older Adolescents	30+	66+	1-2 tablespoons <sup>i</sup>

### How to Give Cornstarch

#### Can be mixed in ...

*Pudding, yogurt, milk, shake, cold juice*

#### Avoid using in...

*Orange juice, formula<sup>ii</sup>*

### Parent Tips

#### Corn Starch Peanut Butter Bars:

*Makes 4 servings*

1/2 Cup Smooth Peanut Butter

1 cup powdered sugar

8 tablespoons Corn Starch

5 tablespoons butter/margarine melted, but wait to mix until it has cooled a little as to not cook the cornstarch

1/2 tsp vanilla

1/2 cup chocolate chips

*Stir powdered sugar and corn starch add to butter vanilla and peanut butter. Place in small pan press down, place in fridge. Melt chocolate chips on very low heat stirring constantly. Spread over top and put back in fridge until it sets. Cut into pieces.*

#### Corn Starch Shake

Vigorously mix cornstarch with 1/2 glass of milk and chocolate syrup and serve before it thickens.

<sup>i</sup> Calculations based on Adult Basal calorie needs per hour of sleep. Assuming basal needs of 50kcal for the first 2-3 hours of sleep. The exact dose will vary based on the boy's weight.

<sup>ii</sup> Will make formula too thick

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