

Complications of Galactosemia

■ Developmental delays

- Cataracts
- Liver disease
- Renal dysfunction
- Hemolytic anemia
- Premature ovarian failure

Bone Mineral Density (BMD)

- Definition: A measure of the amount of minerals, particularly calcium, contained in a certain volume of bone.
- Also known as bone density or bone mass.

Bone Mineral Density

- Calcium gives bones their strength.
- It is constantly being added to and removed from bone as part of the growth process.
- when calcium is removed faster than it is added, bones become lighter, less dense, and more
- * This makes bones weaker and more susceptible to fractures.
- BMD measurements allow us to detect these changes.

Bone Mineral Density

World Health Organization Classification of Bone Mineral Density:

Normal

Osteopenic: Low bone mass.

Osteporotic: Very low bone mass. Severe Osteoporosis: Fractures in addition to

very low bone mass.

What happens in galactosemia?

- Studies have found diminished BMD. Despite appropriate dietary management. Occurs in both sexes.
- Diminished density may result in osteopenia, osteoporosis, frequent bone fractures, and abnormal growth.

Bone Mineral Density

- Factors influencing BMD include physical activity hypogonadism osteocalcin levels nutrition and vitamin status Vitamin K
- Particularly low in females with premature ovarian failure.

Estrogen protects bones from breaking down. Ovarian failure results in low estrogen levels.

Osteocalcin

- Regulatory protein that binds calcium and promotes its addition to bone.
- Activated by a reaction that utilizes vitamin K as a cofactor.
- The ratio of activated to inactivated osteocalcin correlates with serum level of vitamin K.

Vitamin K

- Found in several foods, including leafy green vegetables and legumes.
- Deficiency results in lower levels of activated osteocalcin and contributes to reduced bone mineral density.

Vitamin K

- Vitamin K levels are commonly reduced in children with galactosemia.
- Exact cause of the deficiency is unknown but is thought to be related to the diet.
- Unlike other factors influencing BMD, the role of vitamin K is only becoming apparent in the past decade and requires further research.

Zinc

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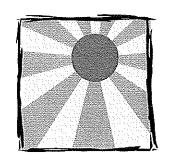
Like vitamin K, zinc is commonly deficient in children with galactosemia.

- Zinc promotes the secretion of Insulin Like Growth Factor-1 (IGF-1) from the liver and enhances receptor responsiveness to the hormone.
- # IGF-1 is vital because it stimulates bone growth and maintains healthy BMD.

Zinc deficiency

- Attributed partially to substitution of animal milk with soy milk, containing phytates.
- Phytates chelate with zinc in the gut and interfere with its proper absorption.
- Absorption can be enhanced by avoiding zinc consumption in close proximity to soy.

Quick Notes - Vitamin D



Smile, the Sun is Shining

Vitamin D plays an important role in bone health by helping us to absorb calcium. It is found naturally in foods like eggs, liver, and fatty fish. Other foods like dairy products and butter are fortified with vitamin D. Most of the fortified foods, you will need to avoid. The good news is that our bodies have the ability to make vitamin D. People with galactosemia who live in Minnesota are at an increased risk for vitamin D deficiency because of the need to avoid fortified dairy products and because of where they live.

Feel the Rays!



During the right time of the year, your body can make enough vitamin D by exposing your hands and face to the sun for 10 to 30 minutes.

Q: Can I visit the tanner to get my vitamin D?

A: Maybe, it depends on the spectrum of wave lengths emitted by the tanning bed. You need wave lengths between 290 to 315 nm.

Q: What time of the year can our bodies make vitamin D?

A: People who live above 40 $^{\rm O}$ N or below 40 $^{\rm O}$ S are not exposed to the right wave length for 3 to 4 months out of the year.

How much Vitamin D do I need?

Just like calcium, your DRI is determined by your age. When you are taking vitamin D supplements, it is possible to take too much. Always check with your doctor or dietitian about how much to take. The body protects itself from making too much vitamin D. Although you may get sun burned by a day at the beach, you won't get an overdose of vitamin D.

Age	DRI (IU/d)
0 - 50 y	200
50 – 70 y	400
> 70 y	600

Quick Notes - Vitamin K

Vitamin K is found in a lot of green leafy vegetables. In addition to this, the bacteria that live in our intestine also make vitamin k. This provides us with about 30 to 50 % of our daily needs.



Food	Serving	Vitamin K (mcg)		
Olive oil	1 Tablespoon	6.6		
Soybean oil	1 Tablespoon	26.1		
Canola oil	1 Tablespoon	19.7		
Mayonnaise	1 Tablespoon	11.9		
Broccoli, cooked	1 cup (chopped)	420		
Kale, raw	1 cup (chopped)	547		
Spinach, raw	1 cup (chopped)	120		
Leaf lettuce, raw	1 cup (shredded)	118		
Swiss chard, raw	1 cup (chopped)	299		
Watercress, raw	1 cup (chopped)	85		
Parsley, raw	1 cup (chopped)	324		

Age	Vitamin K	
	(mcg/d)	
0-6 mo	2	
7 – 12 mo	2.5	
1-3 y	30	
$4-8 \mathrm{y}$	55	
9 - 13 y	60	
14 – 18 y	75	
Males $> 19 \text{ y}$	120	
Females > 19 y	90	

ZINC

ZINC CONTENT OF SELECTED COMMON FOODS

Food (3 ounces, cooked, lean only)		Zinc (mg)
Beef	Shank crosscuts, simmered	8.9
	Chuck, arm pot roast, braised	7.4
	Roundtip, roasted	6.0
	Sirloin, broiled	5.5
	Top round, broiled	4.7
	Ground, 17% fat, broiled	4.6
	Top Ioin, broiled	4.4
	Tenderloin, roasted	4.1
	Eye round, roasted	4.0
Pork	Shoulder, blade, Boston roasted	3.6
	Ham, boneless, 5% fat	2.4
	Tenderloin, roasted	2.2
	Loin chop, broiled	2.0
Lamb	Leg, shank half, roasted	4.3
	Loin, roasted	3.5
Veal	Sirloin, braised	4.0
	Cutlet, pan fried	2.4
Chicken	Liver, simmered	3.7
	Dark meat, roasted	2.4
	Light meat, roasted	1.0
Turkey	Dark meat, roasted	3.8
	Light meat, roasted	1.7
Fish	Tuna, light meat, canned	.7
	Ocean perch, dry heat	.5
	Halibut, dry heat	.5
	Salmon, sockeye, dry heat	.4
Shellfish	Oysters, 6 medium, raw	49.8
	Crab, Alaskan king, moist heat	6.5
	Shrimp, moist heat	1.3

Food		Zinc (mg)	
Dairy	Yogurt, lowfat, plain, 1 cup	2.2	
Products	ducts Milk, lowfat, 1 cup		
	Cheese, cheddar, 1 ounce	.9	
	Cheese, cottage, lowfat, ½ cup	.4	
Cereals	Raisin bran, dry, 1 cup	3.0	
	Shredded wheat, dry, 1 cup	1.0	
	Oatmeal, instant, ½ cup	.6	
	Cream of wheat, instant, ½ cup	.2	
	Corn flakes, dry, 1 cup	.2	
Grains	Bran muffin, 1 medium	.6	
	Brown rice, cooked, ½ cup	.6	
	Bagel, 1-3½ inch	.6	
	Whole wheat bread, 1 slice	.5	
	White rice (enrich), cooked, ½ cup	.4	
	White bread (enrich), 1 slice	.2	
Fruits	Banana, 1 medium	.2	
	Apricots, dried, 7 halves	.2	
	Prunes, dried, 3 medium	.1	
	Orange, 1 medium	.1	
	Apple, 1 medium	.1	
	Raisins, 2 tablespoons	.1	
Vegetables	Peas, green, cooked, ½ cup	.9	
	Potato, baked w/skin, 1 medium	.6	
	Corn, cooked, ½ cup	.4	
	Broccoli, raw, ½ cup	.2	
	Spinach, raw, 1 cup	.2	
	Lettuce, iceberg, chopped, 1 cup	.1	
	Carrots, raw, 1 medium	.1	
Beans/	Baked beans, canned, plain, ½ cup	1.8	
Legumes	Chickpeas, boiled, ½ cup	1.3	
	Kidney beans, boiled, ½ cup	.9	
Meat	Egg, substitute, ½ cup	1.6	
Substitutes	Tofu, raw, ½ cup	1.0	
	Peanut butter, 2 tablespoons	.9	
	Egg, whole, cooked, scrambled	.6	

Quick Notes - Calcium

How much calcium do I need?

Calcium needs vary based upon your age. Just because you have galactosemia, doesn't mean that you need more or less. Look below for your needs.

Age	DRI (mg/d)
0 – 6 mo	210
7 - 12 mo	270
1 -3 y	500
4 - 8 y	800
9 -18 y	1300
19 – 50 y	1000

DRI = dietary reference intake

A closer look at labels

If a product contains calcium, they usually include it on the label. But the label only tells you the percent daily value (%DV). It is easy to translate that into mg of calcium in you know the trick.

- 1. Locate the % DV
- 2. Remove the % sign
- 3. Add a zero
- 4. That is the mg of calcium per serving

** This only works with calcium **



Since I can't eat dairy, how am I supposed to get enough calcium?

Calcium is found in a variety of foods naturally, and more and more products are being fortified. Keep a record of what you eat and see if you are close to your goal.



Cereal, ready to eat	1 cup	100 – 120 mg
Juice, fortified	1 cup	200 – 300 mg
Rice or soy milk, fortified	1 cup	300 mg
Sardines with their bones	2	95 mg
Spinach, cooked	½ cup	125 mg
Beans, cooked	½ cup	45 – 90 mg
Crab	3 oz	50 - 90 mg
Orange	1	50 mg
Broccoli, cooked	1 cup	50 mg
Sweet potato, cooked	1 cup	56 mg
Nuts	1 oz	70
Rolls / hamburger bun	1	60
Lettuce	½ cup	10 mg

Calcium Questionnaire

How often do you eat:	My serving	More than 2 times	1 – 2 times a	2-3 times a	Once a week
	size is:	a day	day	week	
Soy Milk					
Tofu					
Rice Milk					
Calcium fortified water					
Fortified juice					
Calcium fortified bread					
Supplements					
I take a calcium supplem	ent Yes	No	'		
Name of supplement a	ınd mg calcium:				
I take it times	a week				
I take a multivitamin supplement Yes No					
Name of supplement:		•			
I take it times a			_		

Supplements

- 1. Look for supplements that match your nutritional needs.
- 2. Do not take more than 500 mg of elemental calcium at a time.
- 3. Read the ingredient list. Avoid any that contain lactose or carrageen.