

# Update on Nutrition and Kennedy's disease

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# Nutrition Concerns for SBMA

- ▶ Fatty Liver
  - ▶ Nutrition Recommendations
  - ▶ Dietary Supplements possibly beneficial
- ▶ Muscle Concerns-
  - ▶ Fatigue/ Physical Activity Limitations
  - ▶ Muscle Cramps
  - ▶ Nutrients
  - ▶ Dietary Supplements
- ▶ Trouble Swallowing

# Risk Factors for Fatty Liver

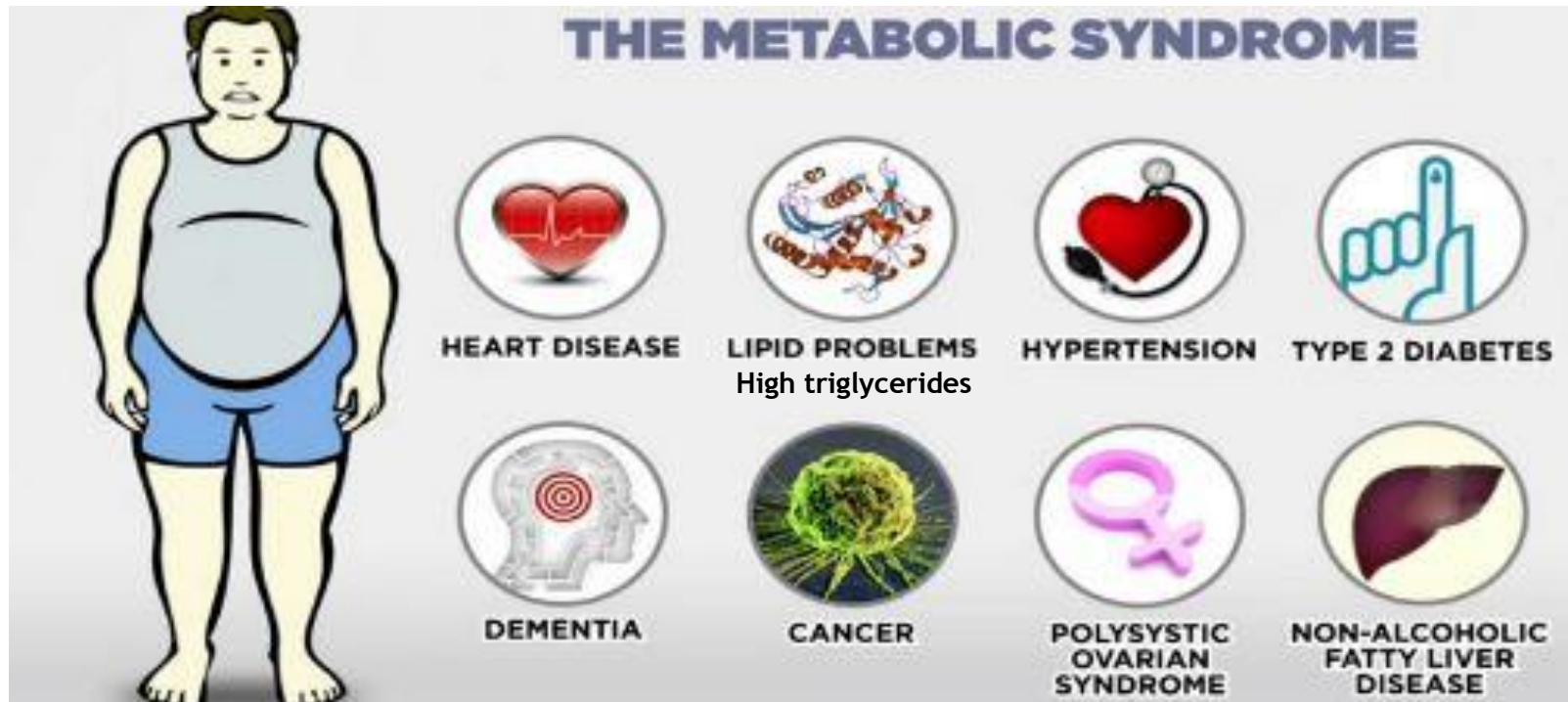
## ▶ Metabolic Syndrome

- Abdominal fat stores
- High Triglycerides ( blood fat)
- High Blood Pressure
- High Blood Sugar

## ▶ High sugar and fat in the diet

## ▶ Limited ability to exercise

# Perhaps some with Kennedy disease have Metabolic Syndrome



# The Metabolic Syndrome

## FOR MEN:

- Waist Circumference  $\geq$  40 Inches
- Triglycerides  $\geq$  150 mg/dL
- HDL Cholesterol  $<$  40 mg/dL
- Blood Pressure  $\geq$  130/85 mm Hg
- Fasting Glucose  $\geq$  100 mg/dL

## FOR WOMEN:

- Waist Circumference  $>$  35 Inches
- Triglycerides  $>$  150 mg/dL
- HDL Cholesterol  $<$  50 mg/dL
- Blood Pressure  $>$  130/85 mm Hg
- Fasting Glucose  $>$  100 mg/dL



# Factors to help prevent Fatty Liver

## 1. Avoid Alcoholic beverages

- Even for those who never drink excessively

Usual alcohol recommendations are

1-2 drinks per day for a man

1 per day for a woman

But even this much alcohol can worsen fatty liver

# Factors to help prevent Fatty Liver

## 2. If you are overweight, commit to losing weight

- Weight watchers or similar respected plan
- Do not follow extremes

Avoiding an entire category of food  
( such as starch) is not appropriate

- Caution- Rapid weight loss  
(more than 1-2 # per week) is bad for your liver

# Factors to help prevent Fatty Liver

## Try to stay within a healthy weight range

Reference Healthy Weights

Body Mass Index ~ 19-25 :

Height ( inches) Weight ( pounds)

▶ 5'	98 to 129 #
▶ 5'2"	104-137#
▶ 5'4"	111-146#
▶ 5'6"	118-155#
▶ 5'8"	126-165#
▶ 5'10"	132-174#
▶ 6'	141-185#

Those with limited activity (sitting most of the day) should weigh 10-20% less than the usual recommended healthy weight for their height

Waist measurements:

- ▶ If  $\geq 35$  inches for women or  $\geq 40$  for men = likely too much abdominal fat.



# Factors to help prevent Fatty Liver

## 3. Limit high sugar foods

Avoid high “doses of sugar”

Too much sugar at once overwhelms the liver  
→ gets stored as fat

- ▶ Sweetened beverages
  - ▶ Regular soda/pop, sweetened coffee drinks, sweet tea  
( **corn syrup especially turns to fat in the liver** )
- ▶ Candy
- ▶ More than 1 serving of fruit at a time
  - ▶ Such as smoothies made with juice,  
several servings of fruit & ice cream
- ▶ Limit sugar/carbohydrate at a meal like those with diabetes should  
( details follow )

# Samples of one carb choice each

**For men choose 4 to 5 “carb choices” per meal.**

**Women - choose 3 to 4 “carb choices” per meal**

## Grains

- 1 slice bread
- ½ large bagel
- ½ cup cooked dried beans
- 1/3 cup pasta or rice
- 1 cup soup
- ¾ cup cold cereal
- ½ cup cooked cereal

## Fruits

- 1 small fresh fruit
- ½ cup fruit
- 1 cup melon or berries
- ½ cup fruit juice
- ¼ cup dried fruit

## Vegetables

- ½ cup potato, corn, peas, corn
- 3 cups raw vegetables
- 1-1/2 cups cooked non-starchy vegetables (i.e. carrots, broccoli...)

## Milk and Yogurt

- 1 cup milk
- 2/3 cup plain or “light” yogurt

## Sweets and Snacks

- ¾ oz. snack food (pretzels, chips, 4-6 crackers)
- 1 oz. sweet snack (2 small sandwich cookies, 5 vanilla wafers)
- 1 Tbs. honey or sugar
- ½ cup ice cream
- 1 cup almond milk ( it is mostly sugar)

# Limit Carbohydrate- ( food that turns into sugar quickly) Do not eliminate it!

For men choose 4 to 5 “carb choices” per meal.

Women - choose 4 “carb choices” per meal

- ▶ 2 small boxes cold cereal = 2 carb choices
- ▶ 8 oz. milk = 1 carb choice
- ▶ ½ cup fruit = 1 carb choice
- ▶ 8 oz. milk = 1 carb choice
- ▶ Egg ( no carbohydrate)
- ▶ Green vegetables - < 1 carb



# Factors to help prevent Fatty Liver

Fatty liver can develop because the body is in a constant state of inflammation.

## 4. Eat anti-inflammatory foods-

- ❑ Eat vegetables, fruits, nuts, seeds, beans, olives, avocado, fish
- ▶ Include colorful foods ( for anti-inflammatory benefits)
- ▶ Include fatty fish ( salmon, sardines, tuna)
- ❑ Eat food with less inflammatory fat - like nuts, seeds, avocado, olives
- ❑ Avoid fried foods, sugary candies, regular soda and salted, fatty meats ( like pepperoni, sausage and bacon)



Photo courtesy of National Cancer Institute

A detailed still life photograph showcasing a variety of nutrient-dense foods. In the foreground, there's a woven basket filled with almonds, a bunch of red radishes, and several bright orange carrots. A yellow squash and a small bag of lentils are also visible. The middle ground features a loaf of braided bread, a halved orange, a head of broccoli, and a small glass bottle of milk. In the background, a jar of pickled vegetables and a whole fish are partially visible. The entire scene is set against a dark, reflective surface, with a green geometric overlay on the right side containing the text.

Eat  
more  
nutrient-  
dense  
foods



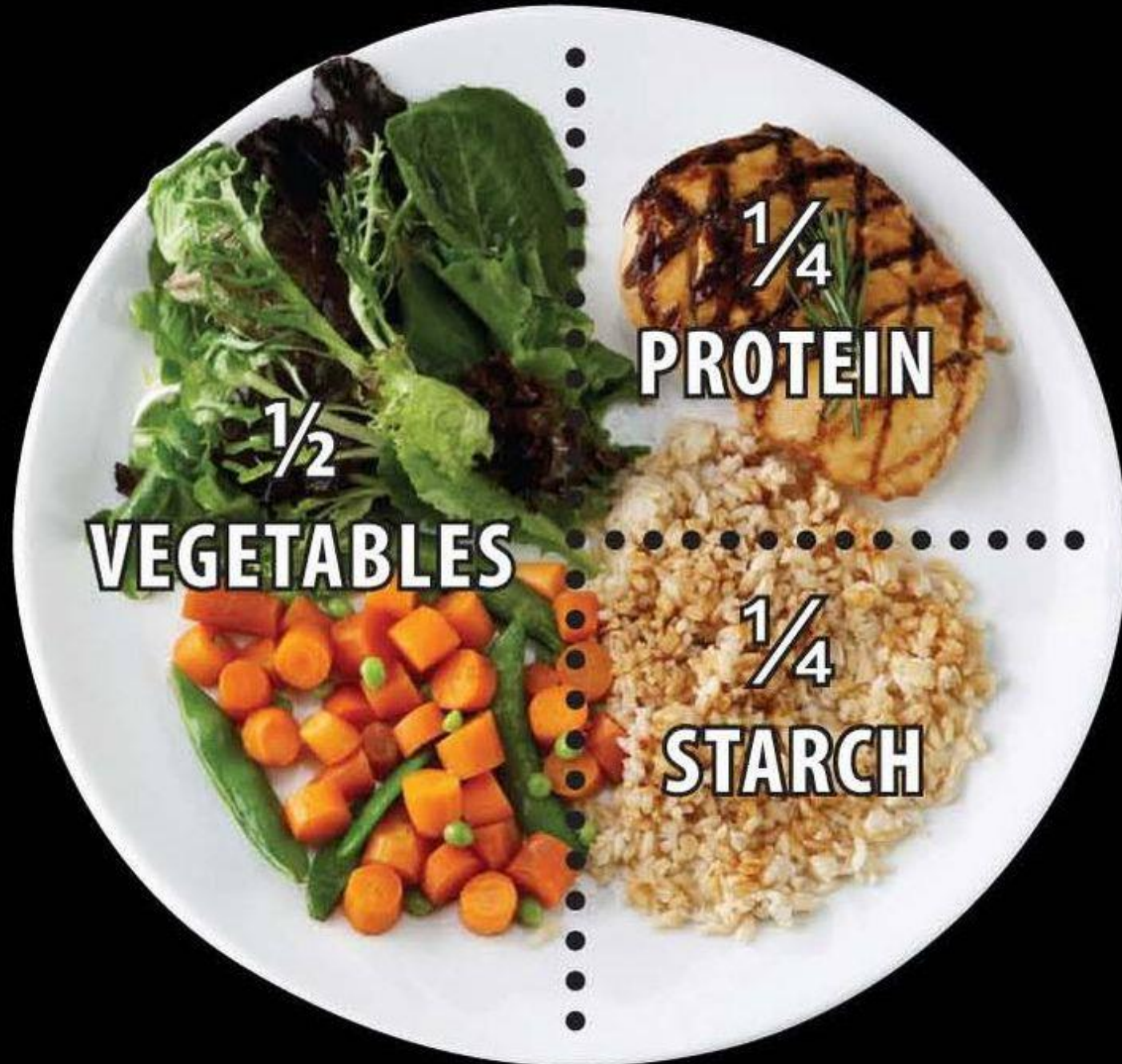


Limit foods  
high in sodium,  
added  
sugars, and refined  
grains



Follow the  
“ My Plate” method

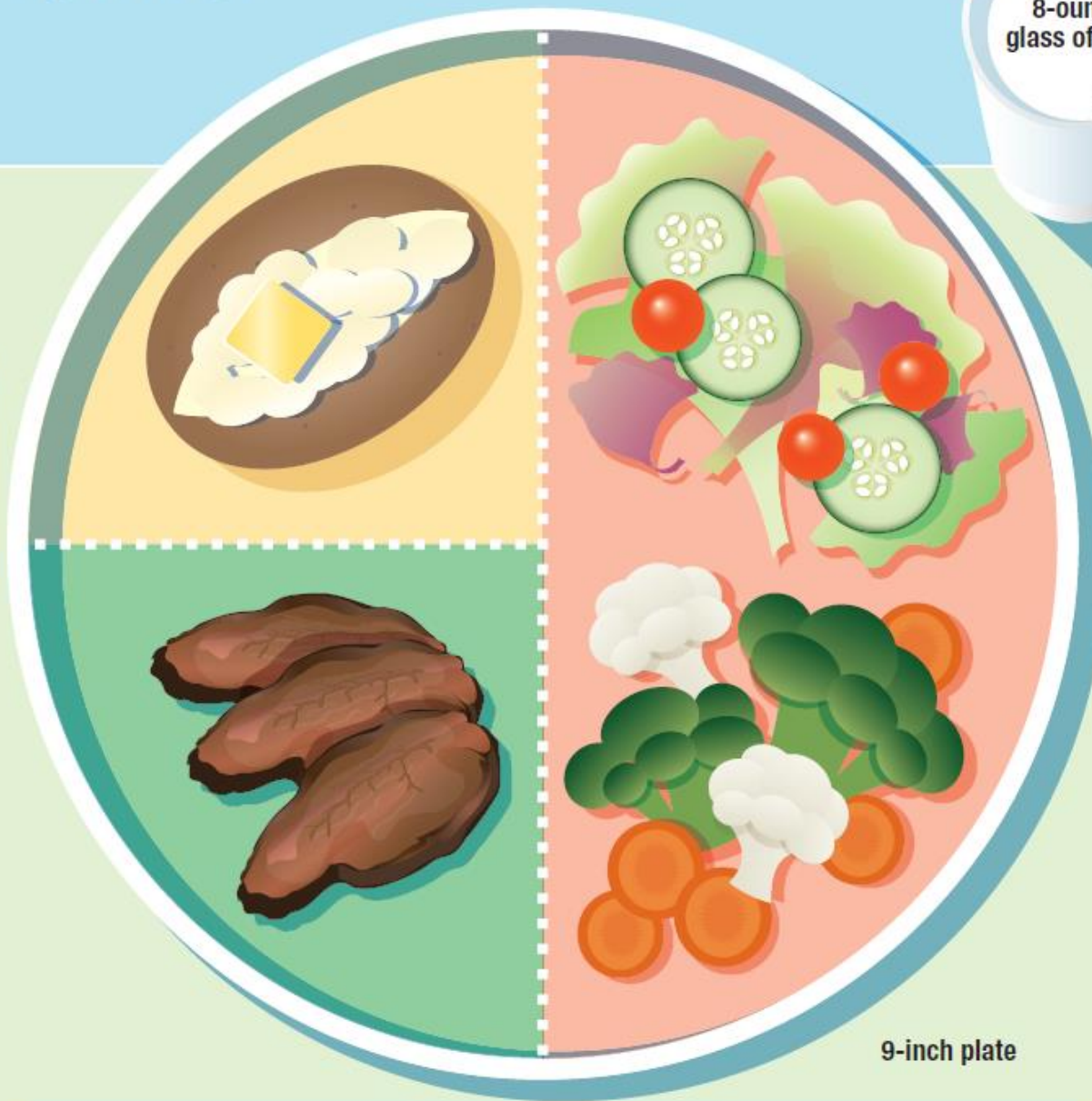
<https://www.choosemyplate.gov/>



# My Plate Planner



Please refer to methods of use, meal planning guidelines, and portion tips on the back of this plate planner.



9-inch plate



Or  
Sugar free yogurt,  
Soy milk,  
cheese stick

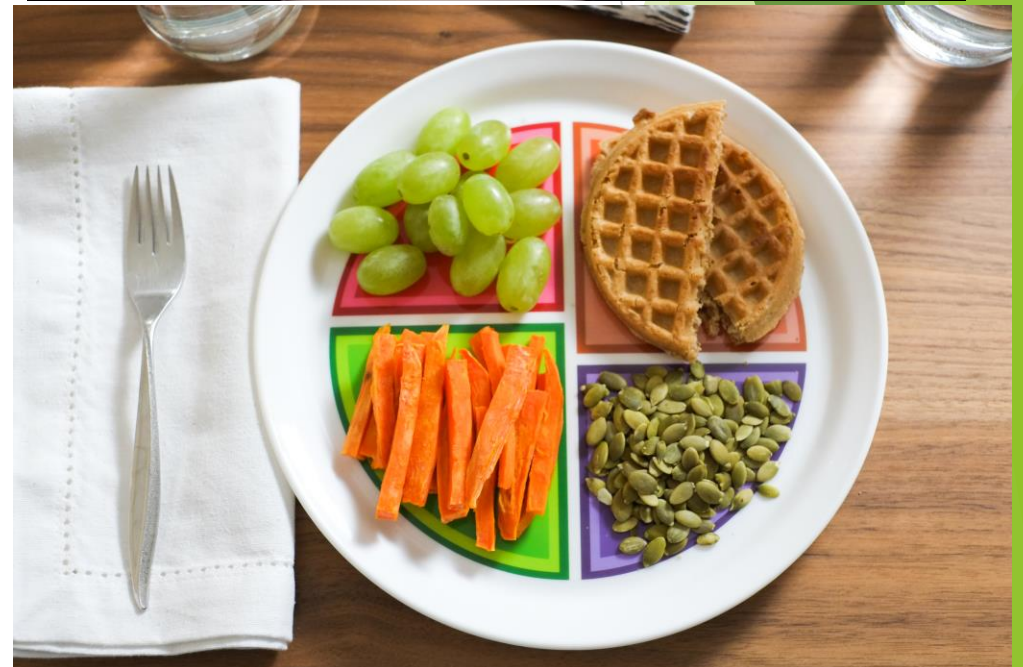
Low calorie  
foods-  
Diet Jell-O,  
flavored  
water







Use the  
“My Plate”  
method  
for  
proportion





**Carbs:** Limit pasta and other carbs to the size of your clenched fist



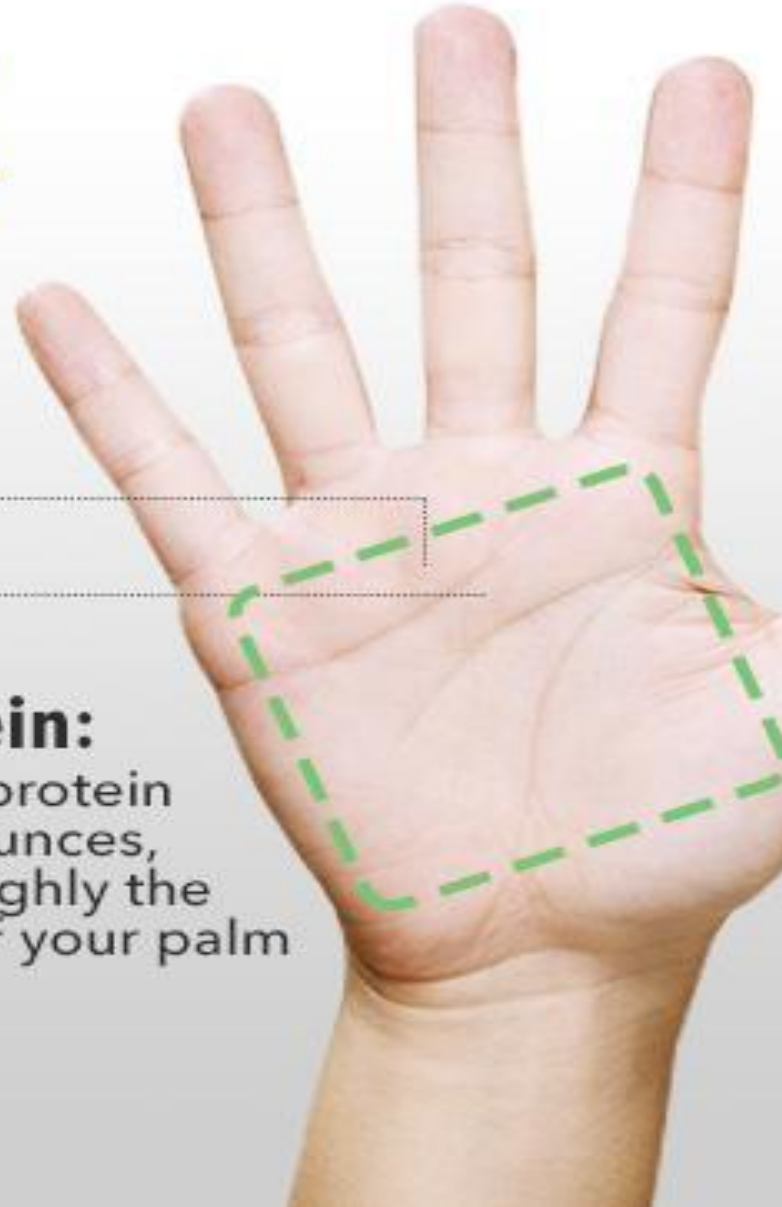
**Fat:** You need a very small amount of fat per day. The size of two thumb tips is all you need.



**Fruit and veg:** You need at least five portions of fruit and veg each day. A portion is as much as you can fit in your cupped hand



**Protein:** Limit protein to 3 ounces, or roughly the size of your palm



# #5. Factors to assure liver health -Eat enough protein



Weight
100 #
125#
150#
175#
200#
225#
250#

Protein Requirement [Grams]

Usual Requirement	Increased with liver disease
37	56
46	69
56	83
65	96
74	110
83	124
93	138







# High Protein Diet - example

3 starches per meal [9 per day]

27 grams

3 grams protein per starch serving:

- ▶ 1 small serving cereal
- ▶ Mini bagel
- ▶ ½ cup noodles/potato/corn

3 oz. protein foods [ twice a day]

42 grams

7 grams protein per ounce

- ▶ Chicken breast [ 3oz.] =21 grams
- ▶ Salmon Steak [small] = 3 oz.

2 cups milk [ soy, dairy] per day

16 grams

8 grams protein per serving

2 servings nuts/seeds

16 grams

- ▶ 2 Tbs. nut butter
- ▶ ¼ cup pumpkin seeds

~100 grams

# Factors to help prevent Fatty Liver-

## 6. Be Cautious of Dietary Supplements

### Avoid iron supplements

- unless advised specifically for you by a doctor aware of all of your lab values ( excess can damage the liver)

### Caution with dietary supplements

- some interact with medicines and many are toxic to the liver
- Ask a pharmacist with access to dietary supplement databases to check for side effects and drug interactions on any supplements you are considering

# Herb Safety

## 1994 Dietary Supplements Health and Education Act -

Vitamins, minerals, herbs = foods.

- ▶ **No Proof of Safety or efficacy needed. Proof of harm falls on the FDA, not the manufacturer.**
- ▶ FDA can't remove a vitamin & mineral supplement considered worthless or offered at dangerously high doses.
- ▶ Can only prevent packages from carrying health claims.
- ▶ Can issue alerts, warnings

# Dietary Supplements- Helpful for Fatty Liver?

## Evidence of Benefit for Fatty Liver:

1. Berberine
2. Choline
3. DHA ( A fish oil)
4. Green Tea
5. Milk Thistle
6. Vitamin E

## ▶ Evidence of Benefit for Other liver Disease- but not for Fatty Liver:

- ▶ SAM-E ( cirrhosis)
- ▶ Fish Oil ( General liver disease)
- ▶ Selenium ( alcohol-related liver disease)



# Dietary Supplements- Helpful for Fatty Liver?

## #1. Berberine

### What is it?

- ▶ A bitter yellow pigment in plants ( Goldenseal, Oregon grape, Barberry)- used in ancient China and India

### Is it Effective?

- ▶ **Insufficient Evidence to Rate Effectiveness for Liver disease.**
  - ❑ Early research suggests that taking berberine for 12 weeks reduces fat in the blood and markers of liver damage in people with liver disease and diabetes.
  - ❑ **Hyperlipidemia** Some preliminary clinical evidence suggests that taking Berberine 500 mg twice daily for 3 months reduces total cholesterol, low-density lipoprotein (LDL) cholesterol, and triglycerides.

### Safety Concerns

- ▶ **Possibly safe** ...when used orally or topically. Berberine has been safely used orally in doses up to 2 grams daily for 8 weeks

- Adapted from *The Natural Medicines Comprehensive Database*-  
<https://naturalmedicines.therapeuticresearch.com/>



# Dietary Supplements- Helpful for Fatty Liver?

## #2. Choline

### What is it?

Choline is nutrient similar to the B vitamins.

It is found in foods such as meats, fish, nuts, beans, vegetables, and eggs. Important for the nervous system. In asthma, choline might help decrease swelling and inflammation.

### Is it Effective?

**Likely Effective for fatty liver disease.** Low blood levels of choline can cause fat to accumulate in the liver.

**Safety Concerns** Adequate intake (AI) of choline for adults is 550 mg per day for males, 425 mg per day for females. Daily Upper Intake Levels (UL) for choline, which is the highest level of intake that is likely to pose no risk of adverse effects, is 3.5 grams for adults.

Doses over 3.5 grams daily are more likely to cause side effects such as sweating, a fishy body odor, diarrhea, and vomiting.



# Dietary Supplements- Helpful for Fatty Liver?

## #3. DHA Docosahexaenoic Acid

### What is it?

- A fat found in cold-water fish ( mackerel, herring, tuna, halibut, salmon) and algae

### Is it Effective? Evidence shows it is possibly effective for:

Liver disease (nonalcoholic fatty liver disease).  
Early research suggests that DHA reduces the risk of severe fat accumulation( pediatric studies).

- Coronary artery disease.
- Lowering Blood fats ( lipids). Research suggests that DHA can lower triglyceride levels

### Safety Concerns

- Likely safe- DHA has been used safely in studies lasting up to 4 years
- Possibly unsafe when used orally in high doses. Doses greater than 3 grams daily might decrease platelet aggregation and increase the risk of bleeding.
  - In women but not men, lower doses of 1 gram daily might have this effect



# Dietary Supplements- Helpful for Fatty Liver?

## #4. Green Tea

### What is it?

Beverage that contains a caffeine type chemical (theophylline) and contains antioxidant polyphenols

### Is it Effective?

Possibly Effective for...

- ▶ Reducing the risk of several types of cancer
- ▶ Reducing risk of Parkinson's disease.
- ▶ Hyperlipidemia
- ▶ Nonalcoholic fatty liver disease

### Safety Concerns

- Possibly unsafe when used long-term in high doses
  - ( fast heart rate, irregular heart rhythm, sleep disturbances)
- Antiplatelet activity-Can prolong bleeding time
- Liver toxicity with high dose polyphenol supplements



# Dietary Supplements- Helpful for Fatty Liver?

## #5. Milk Thistle

**What is it?** A thistle native to Europe brought to America by colonists - now a “weed” throughout the U.S.

**Is it Effective?**

► **Evidence of benefit for :**

- **Diabetes type 2-** lower fasting blood glucose, hemoglobin A<sub>1c</sub> (HbA<sub>1c</sub>), total cholesterol, low-density lipoprotein (LDL) cholesterol, and triglycerides
- **Benign prostatic hyperplasia-** improved lower urinary tract symptoms, urinary flow rates, and residual volumes in men aged 45 to 70 years with BPH
- **Nerve and brain effects:** There is evidence that milk thistle extract protect rat hippocampal neurons against oxidative stress-induced cell death

**Insufficient evidence for benefit in non alcoholic fatty liver disease-**

- Some studies showed possible improvement , most other studies showed no improvement and a large review of studies ( meta-analysis) concluded that **milk thistle lacks a significant effect on mortality or liver disease complications.**

**Safety Concerns**

- **Likely safe** when used orally and appropriately. A specific milk thistle extract in Germany seems to be safe when used in doses up to 420 mg daily for up to 4 years
- **Potential drug interactions-** **should not be used by people taking diabetes medications, statins, warfarin and many other drugs metabolized by the liver** - antipsychotics, beta-blockers such as propranolol (Inderal), metoprolol (Lopressor, Toprol XL), and carvedilol (Coreg).





# Dietary Supplements- Helpful for Fatty Liver?

## #6. Vitamin E



**What is it?** Vitamin E is a fat-soluble vitamin. It is naturally occurring in many foods including vegetable oils, cereal grains, animal fats, meat, poultry, eggs, fruits, and vegetables

**Is it Effective?**

**Possibly effective for Nonalcoholic steatohepatitis (NASH).**

800 IU Vitamin E daily for 24 months significantly improved measures of liver function -liver enzymes, hepatic fibrosis( scarring) , hepatic steatosis ( fat in the liver) , and inflammation

### Safety Concerns

- ▶ **LIKELY SAFE** Likely safe, even at doses exceeding the recommended dietary allowance (RDA. The tolerable upper intake level (UL) in healthy people is 1000 mg/day, equivalent to 1100 IU of synthetic vitamin E or 1500 IU of natural vitamin E
- ▶ **However- it is recommended that non-healthy patients limit Vit E to 400 IU/day or lower.** There is concern that these patients who take doses of 400 IU/day or more might have an increased risk of adverse outcomes and increased mortality from all causes

# Muscle Concerns-

## Muscle Cramps-

- ▶ Electrolytes, fluid

## Muscle Function- Dietary Supplements/Nutrients-

- ▶ Adequate protein
- ▶ Vitamin D
- ▶ Arginine
- ▶ Creatine
- ▶ HMB



# Muscle Cramps- possible causes



## Dehydration

- ▶ Dehydration can interrupt the water balance and increase the amount of electrolytes lost within your muscles.
- ▶ Try to consume enough fluid in the day (enough to produce pale, frequent urine).
- ▶ Most men need more than 12 (eight ounce) cups of fluid per day
- ▶ Most women need 8 (eight ounce) cups of fluid or more per day.
- ▶ Anything “wet at room temperature” counts as a fluid (gelatin desserts, soup, juice, coffee, milk...).

## Low minerals in diet

- ▶ The minerals calcium, sodium, magnesium, and potassium play an important role in contraction and relaxation of a muscle.



# Foods Vs. Supplements

## Superior absorption from food

- i.e. milk protein (casein), lactose & Vit D improve Calcium absorption from milk

## Supplements Compete for absorption –

- i.e. Zinc, Calcium & Iron supplements

## Potential Toxicity from supplements

- There is no toxicity from high intake of the same nutrients from food (i.e. calcium, folic acid, Vit E, B-carotene)
- Food sources don't cause side effects like pill forms can (cramping, diarrhea, nausea), but you may require higher doses than food alone can provide



# Muscle Cramps- possible causes

Try to include rich food sources of these minerals

Nutrient	Sources
Calcium	Milk products, fish with small bones, dark green leafy vegetables, dried beans
Magnesium	Nuts, beans, whole grains, raw green leafy vegetables, soy foods
Phosphorus	Meats, poultry, seafood, milk, yogurt, cheese, tofu
Potassium	Bread, cereal, yogurt, milk, meat, poultry, fish, vegetables, fruits,
Zinc	Seafood, meat, beans, liver, nuts, seeds, poultry, brewer's yeast, whole grains



# Optimal Muscle Function

- ▶ Muscle Training considering fatigue and other limitations ( physical therapists are the experts)

## Nutrients/ Supplements of interest

- ▶ Adequate Protein ( earlier slides)
- ▶ Wide range of minerals from the diet
- ▶ Adequate Vitamin D
- ▶ Arginine
- ▶ Creatine
- ▶ HMB



# Vitamin D

**Source is the sun on your skin ( or supplements)-**

- Food is an insignificant source ( fish, mushrooms)
- Sun on skin can create 10,000 IU (<20 mins.) when you are young -
- We get less efficient at making Vit D as we age

**Vitamin D is important for normal muscle function/balance**

- The darker your skin, higher your fat stores and farther you live from the equator, the greater your risk of deficiency
- For optimal Immunity, muscle function, blood sugar and blood pressure control- assure that your Vitamin D is adequate

**To Test Your Vit D status: 25-OH Vit D**

- Goal reference range > 32 ng/mL or > 80 nmol/L

**To treat deficiency~ 50,000 IU Vit D weekly is usually recommended by Endocrinologists, for months - until levels are normal - then 5,000 IU/day**



# Dietary Supplements- Helpful for Muscle?

## Arginine



**What is it?** An amino acid ( protein building block) - Almonds are a rich source

- ▶ **Is it Effective?** Possibly Effective for:
- ▶ **Erectile dysfunction (ED).** Taking 5 grams of L-arginine by mouth daily seems to improve sexual function in men with ED.
- ▶ **High blood pressure.** There is early evidence that taking L-arginine by mouth can reduce blood pressure in healthy people, people with high blood pressure, and people with slightly high blood pressure with or without diabetes.
- ▶ **Leg pain associated with poor blood flow (peripheral arterial disease).** Research suggests that taking L-arginine by mouth or intravenously (by IV) for up to 8 weeks increases blood flow in people with peripheral arterial disease. However, long-term use (up to 6 months) does not improve walking speed or distance in people with peripheral arterial disease.

### Safety Concerns

- ▶ **Possibly Safe** when used orally and appropriately. L-arginine has been used safely and has only been associated with minor side effects in clinical studies lasting a few days up to 18 months



# Creatine

## What is it?

- ▶ Creatine is a chemical that is found in muscles and the brain.
- ▶ It is also found in foods such as red meat and seafood.

## Is it Effective? Yes, Possibly Effective for:

- Muscle strength
- Athletic Performance -Clinical research on creatine has shown positive results for some measures of athletic performance and mixed results for others

## Not Effective for ALS ( Lou Gehrig's disease) - temporary improvement in muscle strength was not maintained

- ❑ **Safety Concerns?**  
Creatine can cause stomach pain, nausea, diarrhea, and muscle cramping.
- ❑ Also, if you are taking creatine, don't exercise in the heat. It might cause you to become dehydrated.
- ❑ Creatine causes muscles to draw water from the rest of your body. Be sure to drink extra water to make up for this.
- ❑ Many people who use creatine gain weight. This is because creatine causes the muscles to hold water, not because it actually builds muscle.
- ❑ POSSIBLY SAFE when taken in doses up to 10 grams daily for up to 5 years.



# Dietary Supplements- Helpful for Muscle?

## Hydroxymethylbutyrate (HMB)



**What is it?** A protein -derived compound (found in egg white)

### Is it Effective?

#### Possibly Effective for:

- ▶ Increasing body weight and muscle ( studied in people with acquired immunodeficiency syndrome (AIDS).
- ▶ High cholesterol. Early research suggests that HMB can lower cholesterol.
- ▶ High blood pressure. Early research suggests that HMB can lower blood pressure

#### Insufficient Evidence to Rate Effectiveness for:

- ▶ Athletic performance. There is conflicting evidence about the usefulness of HMB for weight training.

### Safety Concerns?

- ▶ Are there any interactions with medications?
- ▶ *It is not known if this product interacts with any medicines.*

*Before taking this product, talk with your health professional if you take any medications.*

# Trouble Swallowing

## ► Speech Language Pathologist

- ❑ Suggestions for chin tucking, etc. can reduce risk of choking
- ❑ If thickened liquids are recommended

- Thickeners with guar gum do not alter the taste of food and do not get thicker as they sit ( like “ Simply Thick”

► If eating becomes a chore, due to the time required to eat a meal, supplemental feeding into the stomach may improve your health and quality of life. Usually you can still enjoy eating- but without the burden of meeting all food, fluid and medicine needs by mouth.

► The Oley Foundation offers contact with other users of “ high tech” nutrition and can be very helpful.



The Oley Foundation  
*Help along the way*  
43 New Scotland Ave., MC-28  
Albany Medical Center  
Albany, NY 12208  
(800) 776-OLEY • [www.oley.org](http://www.oley.org)

**For people fed by tube or IV**





## Dietary Supplement INFORMATION RESOURCES

- ▶ The Natural Medicines Comprehensive database  
<http://www.naturaldatabase.com/>
- ▶ American Botanical Council /  
The Herb Research Foundation <http://www.herbalgram.org/>
- ▶ FDA Dietary Supplements  
<http://www.cfsan.fda.gov/~dms/supplmnt.html>
- ▶ Product Quality Information: Consumer Lab  
<http://www.consumerlab.com>



<http://nccam.nih.gov>

**National library of medicine:**

<http://www.ncbi.nlm.nih.gov>

**NLM Medline plus (for consumers):**

[http://www.nlm.nih.gov/medlineplus/druginfo/herb\\_All.html](http://www.nlm.nih.gov/medlineplus/druginfo/herb_All.html)



**Office of Dietary Supplements** <http://ods.od.nih.gov>

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