Nutrition in ALS:
How to EAT MORE!

Presented by:
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Meet Today’s Expert Panelists

Edward J. Kasarskis, M.D., Ph.D.

Anne-Marie A. Wills, M.D., M.P.H.

Jamie Garry M.S., R.D.
Weight Loss and ALS

- **Body Mass Index (BMI)** is a measure of weight/height.

- **Being underweight** (BMI < 19) is associated with reduced survival in ALS.

Desport, *Neurology* 1999
Increasing calories can improve survival

Hypercaloric enteral nutrition in patients with amyotrophic lateral sclerosis: a randomised, double-blind, placebo-controlled phase 2 trial

Anne-Marie Wills, Jane Hubbard, Eric A Macklin, Jonathan Glass, Rup Tandan, Ericka P Simpson, Benjamin Brooks, Deborah Gellinas, Hiroshi Mitsumoto, Tahseen Mozaffar, Gregory PHanes, Shafeeq S Ladha, Terry Heiman-Patterson, Jonathan Katz, Jau-Shin Lou, Katy Mahoney, Daniela Grasso, Robert Lawson, Hong Yu, Merit Cudkowicz, for the MDA Clinical Research Network

<table>
<thead>
<tr>
<th>Number at risk</th>
<th>Months from randomisation</th>
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<tr>
<td>Control</td>
<td>7  6  5  4  4  3  0</td>
</tr>
<tr>
<td>HC/HC</td>
<td>9  8  8  8  8  8  3</td>
</tr>
<tr>
<td>HF/HC</td>
<td>8  6  6  6  6  3  0</td>
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P=0.03
Goal of Nutritional Intervention in ALS

Maintain Energy Balance

Energy (Food) Intake ≡ TDEE
TDEE = RMR + TEF + EEact
How Can Changes in ALS Status Influence Nutritional (Caloric) Needs?

- **Muscle Atrophy**
  - Changes body composition (loss of lean body mass—skeletal muscle, other tissues and organs) → Reduces Resting Metabolic Rate (calories)

- **Progressive Limb Weakness**
  - Decreased physical activity → Reduces caloric expenditure
  - Decreases ability to feed oneself → Reduces caloric intake

- **Progressive Bulbar Weakness**
  - Dysphagia → Longer meal times → Reduces caloric intake

- **Non-functional Movements**
  - Fasciculations, spasticity, cramps → Increases caloric expenditure
Metabolic Consequences of Inadequate Energy Intake

**Adequate Energy Intake**
- Replete glycogen stores
- Excess calories stored as fat
- Balanced muscle turnover

**Inadequate Energy Intake**
(Fasting, Starvation)
- Depleted glycogen stores
- Fat breakdown for energy
- Muscle breakdown for energy
Two Types of Muscle Atrophy in ALS

- **Neurogenic muscle atrophy**
  - Caused by degeneration of spinal motor neurons and denervation

- **Nutritional muscle atrophy**
  - Caused by inadequate caloric intake vis a vis energy expenditure
How do we estimate calorie needs?

- TDEE using DLW was available from 249 independent determinations in 80 subjects.
- Mean number of observations per subject as $3.2 \pm 1.2$.
- 82.5% of subjects had $\geq 2$ sequential measurements of TDEE.
Kasarskis Equation

- [https://mednet.mc.uky.edu/ALS/](https://mednet.mc.uky.edu/ALS/)

- Men \((66 + (13.7 \times \text{weight}) + (5 \times \text{height}) - (6.76 \times \text{age})) + (55.96 \times \text{ALSFRS6 score}) - 168\)

- Women \((665 + (9.6 \times \text{weight}) + (1.8 \times \text{height}) - (4.7 \times \text{age})) + 55.96 \times \text{ALSFRS6 score} - 168\)
  - Weight in kg
  - Height in cm
  - Age in years
  - ALSFRS-6 (cumulative score of 6 questions)

Estimate daily energy calorie expenditure in ALS using the Harris-Benedict equations and the ALSFRS-6.

A single account can be created by the individual ALS user. The information can be updated by the individual. This way a user can track their calorie and liquid needs each month.

Login & Track Values  Use Calculator

Original research performed at these ALS centers

University of Kentucky
College of Medicine

The University of Vermont
Larner College of Medicine
PennState College of Medicir
The University of Utah
Columbia University
ALS Calculator

Weight

Height

Sex

Choose Sex

Age

Speech

Normal speech processes

Handwriting (with dominant hand prior to ALS onset)

Normal

Dressing and Hygiene

Normal function

Turning in Bed/Adjusting Bed Clothes

Normal

Walking

Normal

Dyspnea

None

Calculate
Example of Kasarskis Calculation

- Age: 60 years old
- Sex: male
- Weight: 180 lbs
- Height: 5’10” (70”)
- ALSFRS-6 Score: Score 19 of 24
  - Speech: 4
  - Handwriting: 2
  - Dressing: 2
  - Turning in Bed: 3
  - Walking: 4
  - Dyspnea: 4
- Harris-Benedict BMR = 1670 kcal
- Adjust for ALS functional status: (55.96 x ALSFRS-6 score) - 168 = 895 kcal
- Total Estimated Daily Caloric Need = 1670 + 895 = 2565 kcal
How can I use this at home?
Electronic-health Application To Monitor Outcomes REMotely (EAT MORE)

- 78 ALS Participants
- MGH ALS Clinic
- Randomized 1:1:1
- In-person RD counseling
- Counseling using mHealth App
- Standard Care
- 6 month dietary intervention
- Outcomes measured at clinic visits
Nutritional counseling with or without mobile health technology: a randomized open-label standard-of-care-controlled trial in ALS

Anne Marie Wills, Jamie Garry, Jane Hubbard, Taylor Mezoian, Christopher T. Breen, Courtney Ortiz-Miller, Paige Nalipinski, Stacey Sullivan, James D. Berry, Merit Cudkowicz, Sabrina Paganoni, James Chan, and Eric A. Macklin

![Graph showing ALSFRS-R change from baseline and percent of recommended calories from baseline for different treatment groups with p-values.](image-url)
ALS Nutrition App

- Nutritional Goals
- Tips to eat more calories
- Recipes
- Monitor your symptoms
  - Screen for swallowing problems
  - Monitor your ALSFRS-R progression
  - Notify providers
- You can choose to share your data with researchers if you want
Disclaimers

- Use of this app does not replace your healthcare providers
- Do not use this app if you are having swallowing difficulties
- If you have a history of diabetes or coronary artery disease (heart attack or stroke) DO NOT use this app. Please consult a registered dietitian.
- If you have allergies to certain foods, do not use recipes which contain those foods.
How to use the App

1. Answer questions about your general health like if you use a feeding tube or have diabetes.

2. If you are willing to participate in research, Consent to sharing your data with researchers.

3. Fill in your age, gender, ALS history, weight, and the ALSFRS-R questionnaire to calculate your new updated diet goals!

-Kasarskis et al., Am J Clin Nutr 2014

Functional Rating

The following questions refer to how you are currently functioning at home.

Please read each item carefully and base your answers on your functioning today compared to the time before you had any symptoms of ALS.

Please choose the answer that best fits your functional status today.

Get Started
Tip of the Day
Add Greek yogurt for extra protein and a creamy texture.

My Calorie Goal
2,532 Calories Per Day

Update Goal In bitesnap

My Data
ALS-FRS-R Score: 40
Current Weekly Weight: 133.0 lbs. (BMI: 22.8)

Recipe Navigator
Use the Recipe Navigator to find recipes by category.

Update Calorie Target
Calories
2226
Submit
Help Me Calculate It
Bitesnap App

- Simply photograph your food
- Bitesnap will recognize what you are eating
- Tell Bitesnap how much you ate
- Bitesnap will calculate how many calories you ate
- Bitesnap will send those calorie results to Apple Healthkit or GoogleFit
Monthly tasks for research only:

- Monitor your ALSFRS-R progression
- Perform tasks for research (tapping test, sustained phonation)
- Screen for swallowing problems
- Prompt you to notify providers about swallowing problems, weight loss
Interested in Using the App?

- We will email out a link to the app to all participants in this webinar
- Patient and caregiver volunteers are needed to
  - Beta test the app
  - Submit Feedback on the app
  - Publicize the app
  - Enroll in the research study using the app
- If interested, please contact:
  - mghALSapp@partners.org
  - Phone: 617-643-2400
Healthy Eating in ALS

Jamie Garry MS, RD, LDN, CBDT
Metabolism & Nutrition Research
Massachusetts General Hospital
Overview

- General nutrition recommendations
- Weight Management Tip
- Common nutrition related problems in ALS
Nutrition Management in ALS

- Higher BMI may increase length and quality of life
  - Avoid weight loss
  - Try to at least maintain weight
  - Weight gain may also be beneficial

- Maintain weight to maintain muscle
- Weight loss can have many causes
Nutrition Management in ALS

- Physician
- Nurse
- Speech-Language Pathologist
- Registered Dietitian
- Physical Therapist
- Occupational Therapist

You
Nutrition Management in ALS
Your Team

- Registered Dietitian (RD)
  - Determine Calorie (energy) and Fluid Needs
  - Complete Body Measurements
    - Height/weight/waist/hip/BMI
  - Provide Appropriate Diet/Recipes Suggestions
  - Assist with Symptom Control
Know Your Numbers

- Simple measurements and calculations
  - Height
  - Weight (invest in a home scale)
  - BMI Calculation – aim for a BMI of >30
    - kg /m²
    - lb/(inches²) 703
  - Online Calculator

- Calorie Needs

http://www.nhlbi.nih.gov/health/educational/lose_wt/BMI/bmicalc.htm
Weight Gain Recommendations

- Aim for slow and steady weight gain
  (0.5 – 1 pound a week)
  ~ extra 250 – 500 calories / day
  3500 calories = 1 pound (rule of thumb)
- Main focus is to avoid weight loss
Weight Gain Recommendations
add 250-500 calories per day

- Eating your calories
- Tube Feed
- Combination of both

Consider a tube feed:
- When ALS interferes with the ability to swallow food and fluids
- When nutrition needs cannot be met by eating
- Before significant weight loss
- Before breathing is severely impaired
General Nutrition Recommendations

Keep it healthy. Choose foods from **all** food groups

- Dairy
- Starches
- Vegetables
- Protein
- Fruit
- Fats/Oils
Easy Ways to Measure out Extra Calories

**Visual Hand Guide to Portion Sizes**

- Your fingertip is about 1 teaspoon
- Your thumb is about 2 tablespoons
- The front of your closed fist is about 1/2 cup
- A clenched fist is the size of a 1 cup serving
- Your palm is 3-4 oz, which is a standard serving of meat

@theflexibledietinglifestyle
What 1800 Calories Looks Like

- **Fruits**
  - 1½ cups
    - 1 cup from the Fruit Group counts as:
      - 1 cup raw, frozen, or cooked/canned fruit; or
      - ½ cup dried fruit; or
      - 1 cup 100% fruit juice

- **Vegetables**
  - 2½ cups
    - 1 cup from the Vegetable Group counts as:
      - 1 cup raw or cooked/canned vegetables; or
      - 2 cups leafy salad greens; or
      - 1 cup 100% vegetable juice

- **Grains**
  - 6 ounces
    - 1 ounce from the Grains Group counts as:
      - 1 slice bread; or
      - 1 ounce ready-to-eat cereal; or
      - ½ cup cooked rice, pasta, or cereal

- **Protein**
  - 5 ounces
    - 1 ounce from the Protein Foods Group counts as:
      - 1 ounce cooked/canned lean meats, poultry, or seafood; or
      - 1 egg; or
      - 1 Tbsp peanut butter; or
      - ¼ cup cooked beans or peas; or
      - ½ ounce nuts or seeds

- **Dairy**
  - 3 cups
    - 1 cup from the Dairy Group counts as:
      - 1 cup milk; or
      - 1 cup yogurt; or
      - 1 cup fortified soy beverage; or
      - 1½ ounces natural cheese or 2 ounces processed cheese

[www.choosemyplate.gov/Resources/MyPlatePlan](http://www.choosemyplate.gov/Resources/MyPlatePlan)
What 1800 Calories Looks Like

Example:

Breakfast
1 cup cooked oatmeal with a banana, and 1 TB of peanut butter, made with milk

Snack:
1 cup plain yogurt with berries

Lunch
2 cups Minestrone soup topped with ~ 1 oz of cheese, served with whole grain crackers

Snack
1 cup baby carrots & snap peas with hummus

Dinner
3 oz salmon with 1 cup brown rice and 1 cup spinach
Gaining Weight the Healthy Way: Choose Heart Healthy Fats

Fat is more calorically dense than protein or carbohydrates so it can be easier to add into one’s diet

- Olive oil, vegetable oil, canola oil
- Avocado
- Olives
- Nuts/ seeds/ nut butter / seed butter
- Fish
- Flax seed/ flax oil

1 TB of oil = ~ 100 calories!
Adding on 100 extra calories a day = 10 pounds a year
Gaining Weight the Healthy Way: High Protein Foods

- Poultry – Chicken, turkey
- Meat – beef, pork, lamb
- Seafood – Fish and shellfish
- Eggs
- Nuts, seeds, nut butters
- Trail mix
- Beans, peas, lentils
- Yogurt
- Greek yogurt has even more protein
- Cottage Cheese
- Cheese
- Milk
  - If you have trouble with milk try alternatives such as soymilk, almond milk, rice milk or coconut milk.
  - Add even more protein and calories by mixing in a tablespoon of dry milk into your milk.
Common Nutrition-Related Problems in ALS

- Decreased appetite
- Fatigue
- Difficulty self-feeding
- Constipation
- Dehydration
- Trouble Swallowing
- Excessive Saliva production
Symptom Management:
Poor Appetite and Fatigue

- Eat when you are hungry
- Eat what appeals to you
- Consume small frequent meals, every 2-3 hours throughout the day
Drink Your Calories

- Drinking your calories can sometimes be easier than eating.
- Try healthier beverage options such as milk (or a milk substitute), 100% fruit juice a smoothie or shake.
- Try commercial supplements like Boost, Ensure, and Carnation-Instant-Breakfast
Symptom Management: Fatigue

- Nap before meals
- Eat soft foods
- Cut food into small pieces
- Choose from all foods groups for more balanced meals
- Avoid foods that require lengthy chewing like steak and salad
- Drink your calories
- Use your feeding tube to supplement your intake
- Accept assistance from friends and family:
  - If someone offers to help if you need anything mention prepared meals would be wonderful!
Symptom Management: Constipation

- **Stay Hydrated**
- **Increase fiber consumption**
  - Whole grain and whole wheat products, such as breads, cereals, pasta, brown rice, oats, barley, and quinoa.
  - Legumes such as cooked dried peas, beans, and lentils.
  - Seeds, such as flax, and chia. Nuts.
  - Vegetables and fruits, including dried.
  - Supplements that contain fiber (Fibersource, Jevity)
- **Discuss with your provider adding in a medication to manage constipation**
Symptom Management: Dehydration

- Water is the best to rehydrate your body, but fruit juices, milk and broth are all good choices.
- Eat foods with high water content such as fruits, vegetables, soups, stews, popsicles, water ice and gelatins.
- Use bottles and cups that are easy to drink from.

- Fluid Needs = 1cc per calorie (rule of thumb)
- 2600 calories = 2600cc/30cc = 86.6 oz/8 oz = 10.8 cups
  - (30cc per fluid ounce)
Symptom Management: Swallowing Difficulty

- Diets with modified textures are a key strategy for people who have difficulty chewing or swallowing.
- Work with your Speech-Language Pathologist to determine the appropriate food texture and beverage thickness, then your dietitian for diet/food suggestions.
- Continue to consume a variety of foods and liquids while following a modified texture diet.
- Modified texture diets change throughout the progression of ALS – reevaluate your needs often!
Symptom Management: Excess Saliva Production

- Remember to swallow frequently, but slowly and carefully.
- Foods that are very sweet or very sour may stimulate salivation.
- Stay well hydrated.
- If you feel that milk thickens your saliva, try soy substitutes for milk, ice cream, and yogurt in drinks and cereal.
- You may be better able to tolerate foods with “cooked” milk such as pudding and custard.
- Eat foods with higher water content such as canned fruits, well cooked vegetables, yogurt, custard and pudding.
Focus on weight maintenance or slow and steady weight gain

Frequent reminders and encouragement help improve nutritional status

Consider early introduction of a feeding tube

Evaluate your nutritional status often

Coordinate Care with Your ALS Team
Thanks!

- Mansi Sharma
- Taylor Mezoian (NCRI)
- Jane Hubbard RD (CRC)
- Jamie Garry RD (CRC)
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- Courtney Ortiz Miller (NCRI)
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