NUTRITION IN ALS

Amyotrophic Lateral Sclerosis
Education day
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SIGNIFICANCE OF NUTRITION IN ALS

- Nutritional status is an independent prognostic factor for survival in ALS patients.\textsuperscript{1, 2, 3, 4, 5}

- Malnutrition is associated with reduced survival\textsuperscript{7} and 7.7 fold increase in mortality risk\textsuperscript{2}
NUTRITIONAL CHALLENGES

• **Energy intake below RDA**: at least 70% of ALS patients ⁸,⁹,¹⁰, ¹¹,¹², ¹³

• **Malnutrition**: 16-55% in ALS patients ⁶

• **Dysphagia in ALS**:
  • more than 81% of patients with advanced ALS ¹⁴
  • 45% of cases of bulbar onset ALS at disease presentation due to an impairment of cranial nerves ¹⁴
Factors contributing to Weight loss & Malnutrition in ALS Patients

- Problem chewing or swallowing food, liquids
- Choking/coughing with meals/ risk of aspiration
- Difficulty with secretions
- Anxiety around mealtime
- Length of time it takes to finish a meal
- Too fatigued and weak to eat
- Unable to prepare meals
- Upper limb weakness and difficulty or inability to feed oneself
- Withholding food or fluids to avoid going to the bathroom
- Constipation
- Dehydration
- Depression
- Decreased appetite
- Difficulty coordinating mealtimes with medications that require spacing of medications and meals/snacks
- Decreased respiratory function
- Hypermetabolism*
NUTRITIONAL CHALLENGES IN ALS

- Specific guidelines for energy, protein and nutrient and fluid requirements have not been established.
- The equations used currently in dietetic practice do not accurately predict the energy needs of an individual ALS patient at different stages of the illness.\textsuperscript{17}
- Hypermetabolism \textsuperscript{6,11, 17, 18}
- Hypometabolism \textsuperscript{19}
NUTRITIONAL MANAGEMENT

- A multidisciplinary approach: Nutritional surveillance in conjunction with the monitoring of the swallowing, respiratory impairment during the disease progression\textsuperscript{16}

- Early nutrition assessment: to optimize time and mode of nutritional intervention\textsuperscript{16} and counselling: on a well-balanced diet high in calories and fat to prevent malnutrition, dehydration, constipation, minimize risk of aspiration and for weight gain/stabilization.
NUTRITIONAL MANAGEMENT

- Ongoing Monitoring of weight (BMI, percentage of weight loss), dietary intake: for any signs of change/deterioration, and to make adjustments in nutritional care plan and counselling.

- Evaluation for timely alternative nutrition support: not meeting nutritional needs, continued weight loss, risk for aspiration.
NUTRITIONAL MANAGEMENT

- Diet Modification based on swallowing capability: to facilitate oral management of food and to reduce risk of aspiration.

**Solid Textures:** soft/moist, cut-up, diced, minced, pureed, blenderized.

**Liquid Consistency:** regular, honey, nectar, puree

*Avoid, dry, crumbly, chewy, sticky textures, mixed textures
NUTRITIONAL MANAGEMENT OF DYSPHAGIA

- **Preparation Methods**: cook food until tender ex: braise, boil, poach, steam. Can mash, puree, or blenderize using food processor. May need strainer or sieve.
- **Puree foods individually so food is identifiable by colour and taste.**

**Thickening products**: commercial products, Resource ThickenUp Clear, Ultra thick. Consiste-Aide, baby cereal
COMMERCIAL PUREED ENTRE

Epikura

Campbell's TresPuree
Nutritional Strategies:\textsuperscript{25}

Nutritional supplements:
Milkshakes, smoothies, Boost 1.5, Ensure 1.5, Resource 2.0, Scandishake, Carnation Instant Breakfast, etc

Increase caloric density of foods: addition of butter, margarine, mayonnaise, cream, oil
Behavioural Strategies:

Smaller, more frequent meals- to reduce fatigue of eating

Visual presentation- to stimulate appetite, give pleasure
  Garnishes- cocoa powder, icing sugar, parsley, lemon wedge even if not eaten.

Sensory stimulation- flavours aroma
**Behavioural Strategies:**

- Maintaining consistent food temperature-hot or cold
  - Keep hot with electric plate warmer or over a dish of hot water
  - Keep liquids cold in chilled insulated cup
- Eat in a quiet controlled environment to reduce distraction, stress, and risk of aspiration.
- If being fed, feeder needs to be aware of appropriate bite size, spoon size, rate of presentation, position of feeder
- Oral hygiene-clear food residue after each meal, snack; clean teeth daily
ALTERNATE NUTRITION (PEG)

The American Academy of Neurology Practice Parameters Update (an evidence-based review)²¹

- PEG should be considered as a nutrition route to stabilize weight in ALS patients with impaired oral food intake.

- PEG should be considered for prolonging survival in patients with ALS.
There are insufficient data to support or refute specific timing of PEG insertion in ALS patients although patients with dysphagia will possibly be exposed to less risk if PEG is placed when FVC is > 50% of predicted.
ALTERNATE NUTRITION (PEG)

The American Academy of Neurology Practice Parameters Update (an evidence-based review)²¹

- There are insufficient data to support or refute PEG for improving quality of life in patients with ALS.

Ultimately patient’s choice whether or not to have PEG
Zhao et al.\textsuperscript{22} (animal study) reported overall significant slower decline in ALS type motor impairment, slower rate of wt loss and increased survival rate compared to control group fed normal diet.

Clinical trial on ALS patients (started December 2009 and estimated study completion December 2012) sponsored by Weill Medical College of Cornell University in collaboration with Johns Hopkins University: Primary outcome measures on safety and tolerability of the ketogenic diet in ALS.
THANK YOU!

QUESTIONS?

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Hyperlipidemia and increased concentrations of circulating apolipoprotein E are positively correlated in survival in ALS. Suggests that increased availability of lipids in blood is beneficial.
References:


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