Nutrition Interventions in Mitochondrial Disorders

AMDF Information Day

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Background

• Role of nutrition is vital in all patients, regardless of diagnosis
• Not a lot of research about how nutrition impacts on MD
• There is no single treatment for MD
  - treatment tailored for each individual
  - lifestyle changes are an effective treatment
• Good nutrition goes hand in hand with pharmaceutical-based treatments eg the “mito cocktail”
Effectiveness of Nutritional Intervention

- Varies from patient to patient; depends on type and severity of disorder
- Generally, those with milder disorders tend to respond better to interventions
- Treatment will not reverse damage already sustained, especially within the brain
- Treatment may be beneficial with instant improvement, or it can take months to see a change
- Benefit of treatment although may not be noticed, but may delay progression of disorder
- Some patients may not benefit from intervention
Symptoms

• Symptoms that may interfere with obtaining good nutrition include:
  - fatigue
  - muscle weakness
  - dysmotility (GI tract does not work properly)
  - dysphagia (inability to swallow)
  - nausea and vomiting
  - ataxia (lack of co-ordination while performing voluntary movements)
  - reflux
Goals of Nutrition Intervention

- Promote normal growth and development
- Promote immune function
- Promote energy production
- Provide sufficient protein, vitamin and minerals
- Preserve and maximise strength, mobility, and functioning – slow the progression of the disease
Energy Loss

- Brain, heart, skeletal muscles, eye muscles and kidneys possibly malfunction because they run out of energy.
- No way to combat this type of energy loss directly:
  - healthy, well-balanced diet is important
  - adequate rest
  - avoid physical stresses eg infections and big temperature changes
  - avoid emotional stressors
Tips for Managing Low Energy

• Frozen/ready to eat meals
• Non-perishable snacks always available
• Oral supplements on hand
• Consider meal replacements
• Snacks when exercising or with extra activity
• Blend foods – easier to tolerate and uses less energy to digest
• Conserve energy – take the lift!
Calories

- *Calories* are a measure of energy in food
- Children with MD often have altered energy needs compared with peers
- Optimising caloric intake has been shown to improve outcomes in some children with MD
- Evaluation of resting metabolic rate may aid in establishing the ideal caloric intake needed for the patient
- Make every mouthful count:
  - offer more energy-dense foods
  - addition of fat or protein to foods
  - remove ‘empty calories’ from diet
  - consider oral supplements
  - consider enteral feeding
Protein

- Protein is stored in the muscles and is needed for strength
- Carbohydrates often easier to eat (and more appealing to children)
- Overloading body with carbohydrates can have a negative effect on the mitochondria
  - mitochondria help break down glucose into energy the cell can use
  - too much glucose can overload the cell in MD
  - protein helps to prevent or minimise glucose peaks
  - complex carbohydrates or low GI carbohydrates also minimise glucose peaks
Protein (cont)

Include protein at each meal and snack

Good sources of protein include:
- meat/poultry/fish
- dairy foods (milk, yoghurt, cheese, custard)
- nuts, beans and legumes
- eggs
- soy products

TIPS
- use nut spreads eg peanut butter, cashew paste on bread
- use skim milk powder in foods/ fluids
- stir cheese through hot foods
- add hummus or tahini to salads
- stir chick peas or beans in meat or vegetable dishes
- add an extra scoop of ice cream or yoghurt to milk drinks
- include eggs at breakfast
- always include a protein serve on a sandwich
Carbohydrate

• *Carbohydrate* is carried in blood and stored in the liver and is needed for energy

• Low vs high glycaemic index (GI)

• Aim for each meal and snack to contain protein AND a low GI carbohydrate where possible and practical
  • allows carbohydrate to be properly and adequately broken down into energy the body can use

**TIPS**

• Swap high GI foods for low GI foods
  • white bread for wholegrain bread
  • pumpkin for sweet potato
  • rice crackers for jatz crackers

• Remove soft drink, cordial, energy drinks

• If having simple sugar foods, combine with low GI food and/or protein
Fluid

- *Hydration* is vital to cell health
- Hydration can be of concern for people with MD
  - physical difficulty taking fluid
  - higher fluid needs

**TIPS**

- Look at urine colour and concentration (B vitamins may discolour urine)
- Determine fluid goal and fill a container at the beginning of day; drink slowly throughout the day
- Use fluids in different forms eg ice confection, dairy desserts, smoothies, milk shakes
- Flavour with small squeeze of fresh fruit juice or diet cordial
Vitamins and Cofactors

• Required for efficient running of the chemical reactions which make energy
• A cofactor can be made by the body, whereas a vitamin cannot, and therefore must be eaten
• For most people, a regular diet contains all the vitamins one could possibly need and their bodies can make as much of any specific cofactor that it needs
• Added vitamins and cofactors may be useful in people with MD
• Because of the varied nature of MD some therapies may be helpful in many, but not in all patients and therefore cannot be considered as "proven and effective"
Vitamins and Cofactors (cont)

• Serve two functions:
  - possibly enhance or stabilise enzyme function and result in improved efficiency of energy generation
  - serve as antioxidants (involved in the prevention of cellular damage) which may slow the progression of the disease

• Examples of antioxidants: vitamin E, coenzyme Q10, lipoic acid, vitamin C, vitamin K and riboflavin (B2)

• Other vitamins and cofactors: creatine, arginine, carnitine, thiamine (B1), folinic acid

• Consult your physician before starting any vitamin or cofactor therapy; over the counter or ‘natural’ doesn’t always mean safe
OTHER CONSIDERATIONS
Poor Appetite

- Often caused by decreased gastric emptying

**TIPS**

- Small, frequent meals
- Higher calorie food options
- Avoid fluids with food
- Avoid big quantities of difficult to digest protein eg animal flesh
- Increased fat can also slow motility
- Look at timing of particular foods/meals
- Medications can be useful
- Consider enteral feeding – overnight feeds, small boluses after meals and snacks
Texture Modified Foods

• Physical deterioration can make it difficult for people with MD to tolerate some textures and thin fluids
  - often require blended foods (soft, mashed, pureed)
  - often require thickened fluids
• Can be as simple as modifying the texture of family meals
• Commercially available thickeners can be added to fluids
• Commercially available thickened products
• Monitor growth
• Monitor fluid intake
Enteral Feeding

• If feeding ability deteriorates, causing faltering growth or growth failure, enteral feeding offers an alternative
• Can provide supplementary or complete nutrition
• Nasogastric tube feeding vs gastrostomy tube feeding
• Consider:
  - Regimen
  - Formula choice
  - Timing of feeds
Dietary Fat Manipulation

Ketogenic Diet
- a high-fat diet that effectively treats some forms of medically refractory epilepsy
- is the standard of care for pyruvate dehydrogenase deficiency, but it is contraindicated in patients with other MDs

Other
- Some patients with MELAS and non-specific MD might benefit from relatively high-fat content in the diet
Exercise

- Exercise can improve mitochondrial functioning
- Where possible, exercise programs should be supervised by a qualified professional
- Young people should start at a very low intensity for brief durations and progress gradually
- Use of a stationary equipment eg exercise bicycle and/or pool therapy may be better tolerated
- A simple carbohydrate–containing drink or meal prior to exercising may increase endurance
- Any exercise program should be halted during illness
Sick Days

- Vomiting and diarrhoea can be a problem
- Dietary treatment temporarily ceased
- Ensure adequate hydration with a glucose and electrolyte solution
- Liaise with treating team
- Amy require admission to hospital for supportive treatment eg IV fluids, tube placement
Sample Meal Plan

**Breakfast**
- Rice bubbles and milk
- White toast with honey
- Orange juice

**Morning Tea**
- Tinned fruit

**Lunch**
- Vegemite white bread roll
- Muesli bar

**Afternoon Tea**
- Jam donut
- Cordial

**Dinner**
- Roast chicken and vegetables
- Soft drink

**Supper**
- Nil

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**Breakfast**
- Oats and milk OR
- Wholegrain toast with peanut butter
- Milo milk

**Morning Tea**
- Tinned fruit stirred in yoghurt

**Lunch**
- Vegemite and cheese spread
- Wholegrain bread roll
- Muesli bar
- Water

**Afternoon Tea**
- Jam donut
- Flavoured milk/fruit smoothie

**Dinner**
- Blended roast chicken and vegetables with gravy
- Water

**Supper**
- Fruit and custard

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Summary

• Plan meals and snacks to include protein and a low GI carbohydrate
• Small, frequent meals are often better tolerated
• Encourage fluid but limit large volumes at meal and snack times
• Modify the textures of foods if necessary to make them easier to eat and digest
• Consider oral supplements or enteral feeds if indicated
• Make the most of meal times when well rested eg eat upon waking; don’t wait until too late in the evening
• Use assistive equipment when feeding
• Remain upright after meals and snacks
• Involve occupation therapists, speech pathologists and exercise physiologists in treatment program

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