Case Study

Library image

Use of peptide feeds to treat gastric complications in Mitochondrial Disease: Lessons learned from a case report

Paula Hynd, Senior Dietitian,
Guisborough Primary Care Hospital, Guisborough

Introduction

Mitochondrial myopathy is a neuromuscular disease caused by damage to the mitochondria. These are small, energy-producing structures that work as the cell's power plant. The cells most affected are nerve cells in the brain and muscle as they require a lot of energy.

The symptoms of mitochondrial myopathy include:

- Muscle weakness (including respiratory muscle) or exercise intolerance
- Heart failure or rhythm disturbances
- Dementia
- Movement disorders
- Stroke-like episodes
- Deafness
- Blindness, droopy eyelids, limited mobility of the eyes, vomiting, and seizures

The prognosis for this disorder ranges in severity from progressive weakness to death. Most mitochondrial myopathies occur before age 20 years, and commence with exercise intolerance or muscle weakness. Muscle cramping, nausea, headache, and breathlessness are also associated with this disorder.

NestléHealthScience

Case Report

In this case, mitochondrial myopathy symptoms in a 64 year old lady resulted in progressive swallowing muscle weakness, breathing difficulties and being wheelchair bound. A puree diet and PEG feed was initially commenced, however due to vomiting and aspiration risk a PEJ tube was inserted. Feed complications were experienced with standard, fibre and an introductory peptide feed. The lady could not tolerate bolus feeding and was therefore fed via an enteral feeding pump. Feed intolerance symptoms included diarrhoea and vomiting.

This resulted in underfeeding as the lady could only tolerate 500ml of feed (Nutrison Peptisorb 2 x 250ml per day at a rate of 80ml/hr x 4-5 hours daily), and was not absorbing some feeds due to vomiting. She lost 3kg over a period of 6 months due to feed intolerance (vomiting and diarrhoea) and lichen planus which resulted in a reduced food intake. This lady's weight in February 2011 was 74kg and in August 2011 was 71kg. At this time a referral was made to speech and language therapy as thick soups were poorly tolerated and pooling occurred at the back of the throat. The speech therapist advised smooth blended thin soups due to aspiration risk. It is worthy of note that the osmolarity of a peptide feed is important when there are intolerance symptoms (diarrhoea and vomiting) and the introductory feed had an osmolarity of 455 mOsmol/litre which is hyperosmolar, and may have worsened symptoms due to this.

On 12/10/2011 a peptide feed with a high MCT fat content (Peptamen®) and lower osmolarity was introduced. Symptoms of vomiting and diarrhoea settled and a feed intake of 1000ml Peptamen® (at a rate of 100ml/hr x 2-3 hours four times per day) was achieved.

On review 11/2011 by telephone the lady reported her weight was maintained at 70kg and she no longer had the adverse feed-related symptoms on a regular basis. On review at home 26/1/12 and 24/2/12 weight remained constant at 70kg and no diarrhoea or vomiting was reported.

Fibre feeds were not tolerated by this lady including standard enteral fibre feeds and Resource® Optifibre® which both resulted in diarrhoea. Regular bowel movements were obtained with regular laxatives.

Conclusion

Although improvements have been made to meet nutritional requirements and treat feed complications, this lady has a progressive illness which will result in ongoing muscle weakness and loss, as she is wheelchair bound and has a very limited exercise capacity due to breathlessness. Adequate nutrition may be important for quality of life, however it will not reduce disease progression. This lady was able to maintain her weight due to a change of peptide feed which she tolerated better.

As her disease progresses there will be further nutritional challenges and regular dietetic review, including adapting volumes, concentrations and types of feed, food and fluids will be required. Ongoing nutritional monitoring using wheelchair scales will be important, as will monitoring of feed intolerance symptoms, quality of life factors, swallowing problems and other medical factors such as development of pressure sores/ nutritional deficiencies. Involvement with the multidisciplinary team will continue to be crucial to detect and treat medical problems which could have an impact on nutrition.

This case study demonstrates that low osmolarity peptide feeds can be safely used to treat feed intolerance in a patient with mitochondrial disease, resulting in weight maintenance and improved quality of life.

Patient Details

Age: 64 **Sex:** F

Diagnosis and Present Complaints:
Mitochondrial myopathy, severely compromised respiratory function, lichen planus.

Previous Medical History:

- Kidney stones.
- Blood transfusion.
- Poor lateral tongue movement and prone to food sticking in her palate which poses a choking hazard.
- Food stuffs lodged in pharynx, pooling and regurgitation/vomiting hence special texture modification advice – food, smooth thin blended and drinks, smooth and via straw.

Social History:
Lives with husband who is retired. Has 2 private carers to do household tasks and prepare and serve meals. Husband administers PEJ feeds.

Medication:
Neomycin, non invasive ventilation overnight and part of the day (BIPAP), breath stacking machine to help with breathing, nebulisers.

Weight History:

- 5/8/2011: 71kg (weighed on wheelchair scales in neurology clinic at a regional specialist centre), height: 1.689m, BMI 24.8kg/m^2.
- 12/10/11: 70kg (BMI 24.5kg/m^2)
- 26/1/12: 70kg
- 24/2/12: 70kg

Usual body weight was 74kg 02/2011, BMI 25.9kg/m^2. Patient felt she was too heavy at this weight and was pleased to lose some weight.

Nutritional Goals:
A target of weight maintenance and minimising feeding complications (vomiting and diarrhoea), in addition to achieving nutritional requirements was set.

Feeding Details:

- Feed name and volume: Peptamen® 4 x 250ml bolus administered via an enteral feeding pump. Provides 1000kcal, 1400ml fluid and 40g protein.
- Route of administration: PEJ (mickey button with jejunal extension).
- Flushes: 50ml cooled boiled water before and after each feed.

Patient Details (cont.)

Feed Tolerance:

- Bowels move using laxatives. Intolerant to Resource® Optifibre® and fibre feeds.
- Cannot tolerate bolus, standard, fibre or high osmolarity peptide feeds due to vomiting and diarrhoea.

Oral Intake:

- Fluid: drinks through a straw Benecol yoghurt x 1 with Actimel x 1 pot per day (and chin tuck technique to prevent aspiration). Sips of tea with milk through a straw, 1-2 cups per day.
- Meals: smooth blended soups, blended mashed potato and gravy or cauliflower and broccoli mashed with cheese sauce, blended trifle.

Nutritional Requirements:

- 1597kcal
- 74g protein
- 2100ml fluid
- 70-100mmol sodium and potassium