



Nutrition and dietary therapy

Many people with Pompe disease have trouble with eating. Muscle weakness can make it hard to bite, chew, or swallow food. If eating becomes an effort, you or your child may not get the nourishment needed to keep up strength and energy. You may lose weight or have trouble keeping it on. Weak muscles may also slow the passage of food through the body. This can lead to digestion or bowel problems. Changing your diet or your eating habits or getting nutrition through tube feeding can help manage these problems. This handout explains the benefits of dietary therapy for people with Pompe disease.



Q Why do people with Pompe disease have trouble with eating?

A As glycogen builds up in the cells, the muscles that we use to eat and digest food get weaker and weaker. This affects people in different ways. Weakness of the face or mouth muscles can make it hard to chew. Weakness of the tongue or throat muscles can make it hard to swallow. Most people with severe muscle weakness try to cope by eating small meals or avoiding certain foods. But some are too tired to eat because of breathing problems that cause fatigue. For others, the desire to eat may be reduced by nighttime breathing problems that disrupt sleep. Whatever the reason, if you cut back too much on what you eat, you may lose weight or become malnourished. This means your body is not getting enough food to keep you healthy. Some people may need a feeding tube to get enough nutrition (see *Why do people with Pompe disease need tube feedings?* on page 3). In less severe cases, there are a number of steps a person can take to get the nutrition the body needs.

Q What steps can I take to improve my diet and eat better?

A Foods that are too large, very thick, or solid can be difficult to chew or swallow. To make them easier to eat, mash them, chop them into smaller pieces, or mix them with some sauce or gravy. Take small mouthfuls and chew food well. Eat several small meals instead of 3 large ones. Be sure to drink enough liquids throughout the day and sip them slowly. Drinking with a straw and staying upright for an hour or 2 after eating may also be helpful. Food and drinks that are thin (such as soup, broth, water, or milk) can also be hard to swallow. They can move through the throat too quickly and make you gag. Try thickening drinks and soups with baby rice cereal, cornstarch, or special powders that are made for this purpose. A dietitian can help you plan well-balanced meals. You may be able to replace meals with nutritional drinks that supply needed vitamins and minerals.

Other names for Pompe disease

Acid alpha-glucosidase deficiency, acid maltase deficiency (AMD), glycogen storage disorder (GSD) type II, glycogenosis II, and lysosomal alpha-glucosidase deficiency. In different parts of the world, Pompe may be pronounced "pom-PAY," "POM-puh," or "pom-PEE."

Q What can I do to manage digestion or bowel problems?

A The best advice is to talk with your healthcare provider. Describe the problems you are having and ask for help in managing the symptoms. Weakness of the muscles that move the food toward the stomach may cause heartburn, or acid reflux (also called GERD). This occurs when swallowed food and stomach acids flow back toward the mouth through the esophagus (the pathway that carries the food from the throat to the stomach or the small intestine). Eating small meals and keeping your head up when you eat may be helpful.

Weakness of the muscles in the chest or abdomen can make it hard to push waste out of the body when you have a bowel movement. This may lead to constipation. Constipation can also lead to diarrhea. Eating a diet high in fiber and drinking lots of fluids can help prevent constipation. Be sure to check with your healthcare provider before taking any medicines for digestion, diarrhea, or bowel problems.

Q I've heard that a high-protein diet can help people with Pompe disease. What's known about it?

A A high-protein diet does seem to help some children and adults with late-onset Pompe disease. The diet is based on the theory that eating more protein and fat (like meat, eggs, cheese, and butter) and fewer carbohydrates (like bread and pasta) may help slow the muscle weakness that occurs when too much glycogen builds up in the cells. There are some patients whose muscle strength and ability to walk have improved after following a high-protein diet. But many others showed no sign of improvement. Be sure to check with your child's doctor before starting an infant on this type of diet. More studies are needed to see if the diet will have positive results for larger numbers of patients.

Q How can alanine and other nutritional supplements help people with Pompe disease?

A Alanine is an amino acid, one of the building blocks of protein. A few studies have suggested that alanine may give children and adults with Pompe disease more energy if taken in small amounts through the day. Though the scientific evidence is limited, healthcare providers who favor this approach feel that alanine supplements help to prevent muscle wasting when glycogen builds up in the cells. Alanine comes in a powdered form that can be mixed with food.

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Why do people with Pompe disease need tube feedings?

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Tube feeding provides complete nutrition for people who are not able to take food by mouth. This may include people with severe swallowing or breathing problems, or babies with infantile-onset disease who are too weak to suck from a breast or bottle or are not gaining weight. Tube feeding also helps keep food from getting trapped in the lungs when it “goes down the wrong way” and ends up in the windpipe, a common problem for people who have trouble swallowing. With tube feeding, also called **enteral nutrition**, liquid food goes into the stomach through a thin plastic tube.

There are 2 types of feeding tubes:

- A **nasal tube** carries the liquid food to the stomach through the nose, throat, and esophagus
- A **gastrostomy tube** (or G-tube) brings food directly into the stomach through an opening in the stomach wall. Surgery is needed to attach the G-tube. It is a good option for people who need a feeding tube for more than a short time

Where to learn more

For more advice and information about dietary therapy for people with Pompe disease, these sources may be helpful:

- The **International Pompe Association (IPA)** can direct you to Pompe disease patient groups around the world. To find the contact for your country, visit the IPA Web site at www.worldpompe.org. The Web site also has links to current research articles on Pompe disease
- **Caring for a child with Pompe’s disease** offers tips on feeding a baby with infantile-onset Pompe disease. It is available online at <http://www.pompe.org.uk/agsdarne.html>
- The **Acid Maltase Deficiency Association (AMDA)** sponsors a telephone conference series on Pompe disease. To review a summary of the presentation on Nutrition and Exercise Therapy, visit the AMDA Web site at <http://www.amda-pompe.org>

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