Ménétrier Disease

What is Ménétrier disease?
Ménétrier disease causes the ridges along the inside of the stomach wall—called rugae—to enlarge, forming giant folds in the lining of the stomach. The rugae enlarge because of an overgrowth of surface mucous cells of the stomach. In a normal stomach, rugae release protein-containing mucus. Enlarged rugae release too much mucus, causing a leakage of protein from the blood into the stomach. This shortage of protein in the blood is known as hypoproteinemia. Ménétrier disease also causes a decrease in stomach acid resulting from a reduction in acid-producing parietal cells.

People with Ménétrier disease suffer from severe stomach pain, nausea, frequent vomiting, and other symptoms. They also have a higher risk of developing stomach cancer, also called gastric cancer.

Ménétrier disease is also called hypoproteinemic hypertrophic gastropathy.

Other conditions that can cause enlarged rugae but are not Ménétrier disease include:
- Zollinger-Ellison syndrome—a condition in which tumors in the pancreas cause the stomach to make too much acid
- syphilis—a type of sexually transmitted bacterial infection
- cytomegalovirus—a type of viral infection
- histoplasmosis—a type of fungal infection
- linitis plastica—a type of gastric cancer
- gastric lymphoma—a type of cancer originating in the stomach

What causes Ménétrier disease?
What causes Ménétrier disease is unclear; however, it is thought to be an acquired disorder with no known genetic component. Recent studies suggest people with Ménétrier disease have stomachs that make abnormally high amounts of a protein called transforming growth factor-alpha (TGF-α). Growth factors are proteins in the body that tell cells what to do, such as grow larger, change shape, or divide to make more cells. A cause for the overproduction of TGF-α has yet to be found.

Who gets Ménétrier disease?
Ménétrier disease is rare and more common in men, usually appearing between the ages of 30 and 60.

What are the symptoms of Ménétrier disease?
Possible signs and symptoms of Ménétrier disease include:
- severe pain in the top middle part of the abdomen
- nausea and frequent vomiting
- swelling of the face, abdomen, limbs, and feet
• loss of appetite
• extreme weight loss
• malnutrition
• low blood protein
• anemia
• diarrhea

How is Ménétrier disease diagnosed?
Ménétrier disease is diagnosed through x rays, endoscopy, and biopsy of stomach tissue. Endoscopy involves looking at the inside of the stomach using a long, lighted tube inserted through the mouth. During a biopsy, the doctor removes a small piece of tissue and examines it with a microscope for signs of disease.

How is Ménétrier disease treated?
Treatment may include medications to relieve nausea and pain. A high-protein diet is prescribed to offset the loss of protein from enlarged rugae. Part or all of the stomach may need to be removed if the disease is severe.

The anticancer drug cetuximab (Erbitux) blocks the action of TGF-α and is being investigated as a promising new treatment for Ménétrier disease.

Points to Remember
• Ménétrier disease causes the ridges along the inside of the stomach wall—called rugae—to enlarge, forming giant folds in the lining of the stomach.
• Ménétrier disease is rare and more common in men, usually appearing between the ages of 30 and 60.
• Recent studies suggest people with Ménétrier disease have stomachs that make abnormally high amounts of transforming growth factor alpha (TGF-α)—a protein that tells cells what to do.
• Ménétrier disease is diagnosed through x rays, endoscopy, and biopsy of stomach tissue.
• Treatment for Ménétrier disease may include medications to relieve nausea and pain and surgery to remove part or all of the stomach.
Hope through Research

The National Institute of Diabetes and Digestive and Kidney Diseases conducts and supports basic and clinical research into many digestive disorders, including Ménétrier disease.

Participants in clinical trials can play a more active role in their own health care, gain access to new research treatments before they are widely available, and help others by contributing to medical research. For information about current studies, visit www.ClinicalTrials.gov.

For More Information

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The National Digestive Diseases Information Clearinghouse (NDDIC) is a service of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). The NIDDK is part of the National Institutes of Health of the U.S. Department of Health and Human Services. Established in 1980, the Clearinghouse provides information about digestive diseases to people with digestive disorders and to their families, health care professionals, and the public. The NDDIC answers inquiries, develops and distributes publications, and works closely with professional and patient organizations and Government agencies to coordinate resources about digestive diseases.

Publications produced by the Clearinghouse are carefully reviewed by both NIDDK scientists and outside experts. This publication was reviewed by Robert J. Coffey Jr., M.D., Vanderbilt University Medical Center.

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