



## MENETRIER'S DISEASE

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### ABSTRACT

**Menetrier disease** is otherwise called as gastropathy, is a rare, acquired, premalignant disease of the stomach characterized by massive gastric folds, excessive mucous production with resultant protein loss, and little or no acid production. The disorder is associated with excessive secretion of transforming growth factor alpha. The untreated cases may turned to cancer stomach.

#### Key words:

Gastropathy, Stomach, Protein, Mal-absorption, Cancer

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## INTRODUCTION

**Menetrier disease** (also known as hypoproteinemic hypertrophic gastropathy; named after a French physician Pierre Eugène Menetrier, 1859–1935), is a rare, acquired, premalignant disease of the stomach characterized by massive gastric folds, excessive mucous production with resultant protein loss, and little or no acid production. The disorder is associated with excessive secretion of transforming growth factor alpha (TGF- $\alpha$ ). Menetrier's disease causes the ridges along the inside of the stomach wall—called rugae—to enlarge, forming giant folds in the stomach lining. The rugae enlarge because of an overgrowth of mucous cells in the stomach wall.



### Differential Diagnosis

Imaging differential considerations for diffuse rugal thickening include

gastric lymphoma / may also occur as part of post transplant lymphoproliferative disorder affecting the stomach.

forms of gastritis

- Infective Gastritis
- Radiation Gastritis
- Eosinophilic Gastritis

### Epidemiology

The average age of onset is 40 to 60 years, and men are affected more often than women. Adults with Menetrier disease have a higher risk of developing gastric adenocarcinoma.

### Causes

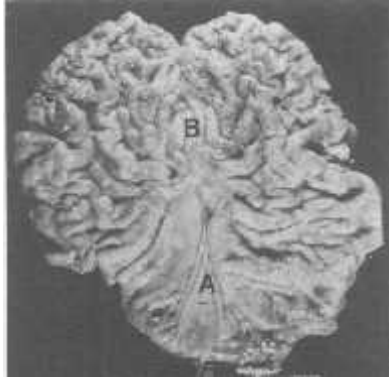
The cause of Menetrier disease is unknown, but it has been associated with HCMV infection in children and H. pylori infections in adults. Additionally, increased TGF- $\alpha$  has been noted in the gastric mucosa of patients with the disease.

Studies suggest that people with Menetrier's disease have stomachs that make abnormally high amounts of a protein called transforming growth factor-alpha (TGF- $\alpha$ ).

### Epidemiology

Rare disease with incidence < 1 per 200,000. Bimodal in distribution, children often < 10 years, adult forms 30-60 years, peak 55 years. Male preponderance both in juvenile and adult form.

## Pathology



The exact aetiology is not well known. The juvenile form has been linked to cytomegaloviral infection and usually resolves spontaneously. The adult form tends to progress with time and TGF- $\alpha$  activation of the EGFR receptor has been implicated.

The characteristic of the disease is gastric mucosal hypertrophy, which may cause the rugae to resemble convolutions of the brain. Rugal thickening is predominantly caused by the expansion of the epithelial cell compartment of the gastric mucosa.

## Location

It most commonly affects the gastric fundal region also any part of the stomach may be involved.

## Signs and Symptoms

The most common symptom of Menetrier's disease is pain in the upper middle part of the abdomen. It includes Achlorhydria, hypoproteinaemia and oedema. Hypoproteinaemia may lead to ascites and pleural effusions. Other signs and symptoms of Menetrier's disease may include

- Nausea and frequent vomiting
- Diarrhea
- Loss of appetite
- Extreme weight loss
- Malnutrition
- Low levels of protein in the blood
- Swelling of the face, abdomen, limbs, and feet due to low levels of protein in the blood
- Anemia—too few red blood cells in the body, which prevents the body from getting enough oxygen—due to bleeding in the stomach

People with Menetrier's disease have a higher chance of developing stomach cancer.

## Diagnosis

**Medical and family history** - Taking a medical and family history is one of the first things a health care provider may do to help diagnose Menetrier's disease. He or she will ask the patient to provide a medical and family history.

**Physical exam** - A physical exam may help diagnose Menetrier's disease. During a physical exam, a health care provider usually

- examines a patient's body
- uses a stethoscope to listen to bodily sounds
- taps on specific areas of the patient's body

**CT scan** - CT scans use a combination of x rays and computer technology to create images. For a CT scan, a health care provider may give the patient a solution to drink and an injection of a special dye, called contrast medium. CT scans require the patient to lie on a table that slides into a tunnel-shaped device where an x-ray technician takes x rays. An x-ray technician performs the procedure in an outpatient center or a hospital, and a radiologist—a doctor who specializes in medical imaging—interprets them. The patient does not need anesthesia. CT scans can show enlarged folds in the stomach wall.

**Upper GI endoscopy** - This procedure involves using an endoscope—a small, flexible tube with a light—to see the upper GI tract, which includes the esophagus, stomach, and duodenum. A gastroenterologist—a doctor who specializes in digestive diseases—performs the test at a hospital or an outpatient center. The gastroenterologist carefully feeds the endoscope down the esophagus and into the stomach. A small camera mounted on the endoscope transmits a video image to a monitor, allowing close examination of the stomach lining. The gastroenterologist also can take a biopsy of the stomach tissue during the endoscopy. A health care provider may give a patient a liquid anesthetic to gargle or may spray anesthetic on the back of the patient's throat. A health care provider will place an intravenous (IV) needle in a vein in the arm to administer sedation. Sedatives help patients stay relaxed and comfortable. The test can show enlarged folds in the stomach wall.

**Biopsy**- Biopsy is a procedure that involves taking a piece of stomach tissue for examination with a microscope. A gastroenterologist performs the biopsy at the time of upper GI endoscopy. A pathologist—a doctor who specializes in diagnosing diseases—examines the stomach tissue in a lab. The test can diagnose Menetrier's disease by showing changes in the stomach's mucous cells and acid-producing cells.

**Blood test** - A health care provider will take a blood sample that can show the presence of infection with *H. pylori* or CMV. A blood test involves drawing blood at a health care provider's office or a commercial facility and sending the sample to a lab for analysis.

## Treatment

Treatment may include medications, IV protein, blood transfusions, and surgery.

## Medications

Health care providers may prescribe the anticancer medication cetuximab (Erbix) to treat Menetrier's disease. Studies have shown that cetuximab blocks the activity of epidermal growth factor receptor and can significantly improve a person's symptoms, as well as decrease the thickness of the stomach wall from the overgrowth of mucous cells. A person receives cetuximab by IV in a health care provider's office or an outpatient center. Studies to assess the effectiveness of cetuximab to treat Menetrier's disease are ongoing. A health care provider also may prescribe medications to relieve nausea and abdominal pain.

In people with Menetrier's disease who also have *H. pylori* or CMV infection, treatment of the infection may improve symptoms. Health care providers prescribe antibiotics to kill *H. pylori*. Antibiotic regimens may differ throughout the world because some strains of *H. pylori* have become resistant to

certain antibiotics—meaning that an antibiotic that once destroyed the bacterium is no longer effective. Health care providers use antiviral medications to treat CMV infection in a person with a weakened immune system in order to prevent a serious disease from developing as a result of CMV. Antiviral medications cannot kill CMV; however, they can slow down the virus reproduction.

#### ***Intravenous Protein and Blood Transfusions***

A health care provider may recommend an IV treatment of protein and a blood transfusion to a person who is malnourished or anemic because of Menetrier's disease. In most cases of children with Menetrier's disease who also have had CMV infection, treatment with protein and a blood transfusion led to a full recovery.

#### ***Surgery***

If a person has severe Menetrier's disease with significant protein loss, a surgeon may need to remove part or all of the stomach in a surgery called gastrectomy.

Surgeons perform gastrectomy in a hospital. The patient will require general anesthesia. Some surgeons perform a gastrectomy through laparoscopic surgery rather than through a wide incision in the abdomen. In laparoscopic surgery, the surgeon uses several smaller incisions and feeds special surgical tools through the incisions to remove the diseased part of the stomach. After gastrectomy, the surgeon may reconstruct the changed portions of the GI tract so that it may continue to function. Usually the surgeon attaches the small intestine to any remaining portion of the stomach or to the esophagus if he or she removed the entire stomach.

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