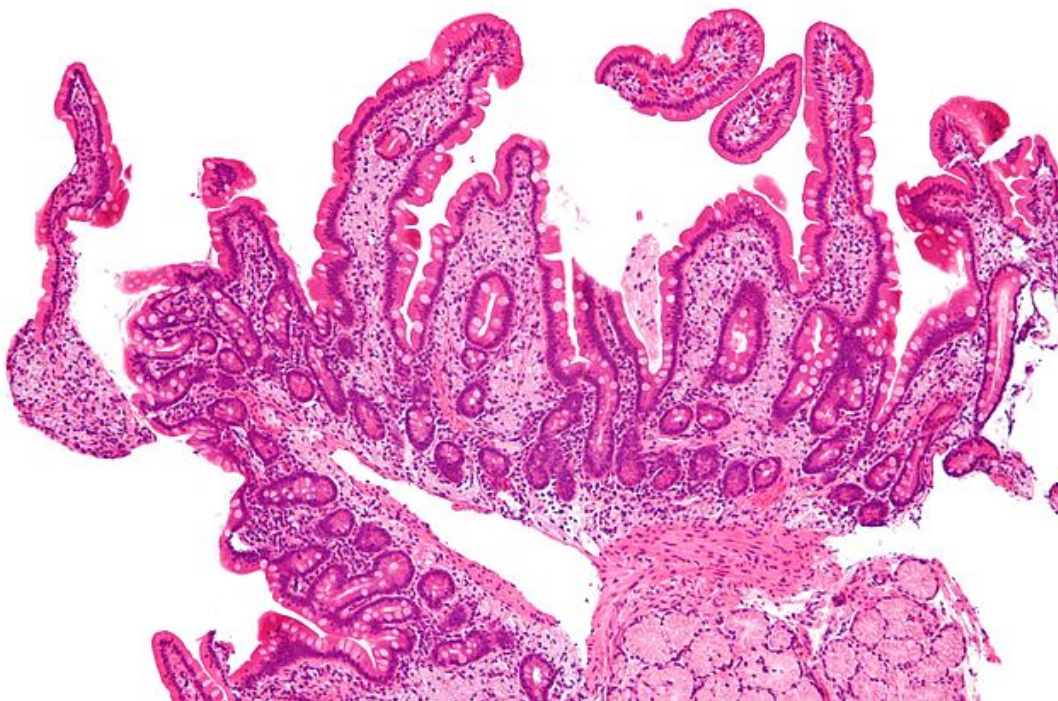


## Whipple's Disease — Symptoms and Diagnosis

[See online here](#)

Whipple's disease is a rare infection affecting several tissues throughout the body including the GI tract, central nervous system, and heart by the bacteria *Tropheryma whippelii*. Symptoms include malabsorption, weight loss, and diarrhea with extra GI symptoms including arthritis and joint pain. The following article contains all important facts about the Whipple's disease.



### Definition of Whipple's Disease

Whipple's disease is named after George Hoyt Whipple who identified it in 1907. It is a rare systemic illness and is caused by **infection of *T. whippelii***. The first symptom of infection is joint pain. Much later [diarrhea](#) and malabsorption follow. Without proper antibiotic treatment, death is likely within one year.

### Epidemiology of Whipple's Disease

There have **only** been about **1000 cases** since described in 1907. Whipple's disease is **found in men much more frequently** than in women and in Caucasians more than any other race.

# Etiology of Whipple's Disease

*Tropheryma whippelii* is a **gram positive Actinobacteria** that can be found intracellularly and extracellularly. The bacteria has been isolated from synovial fluid, CSF, and lung biopsies. It grows in the acidic vacuoles of cells. This may increase its virulence and penchant for chronic infection. The bacteria may be part of many people's natural flora and only becomes a problem when the body starts reacting to it. Alternatively, the bacteria are also found in soil and on both wild and domestic animals.

## Pathology and Pathophysiology of Whipple's Disease

*Tropheryma whippelii* infiltrates tissue and causes destruction. The **inflammatory process** that follows is also disruptive and **damages tissues**. As part of the immune response, macrophages absorb the bacteria. There is some research that implies a predisposition to Whipple's disease in patients that have difficulty breaking down intracellular bacteria. In the GI tract, especially in the duodenum and [small intestine](#), Whipple's disease **causes malabsorption by infiltrating the mucosa** down to the lamina propria and disrupting villous function.

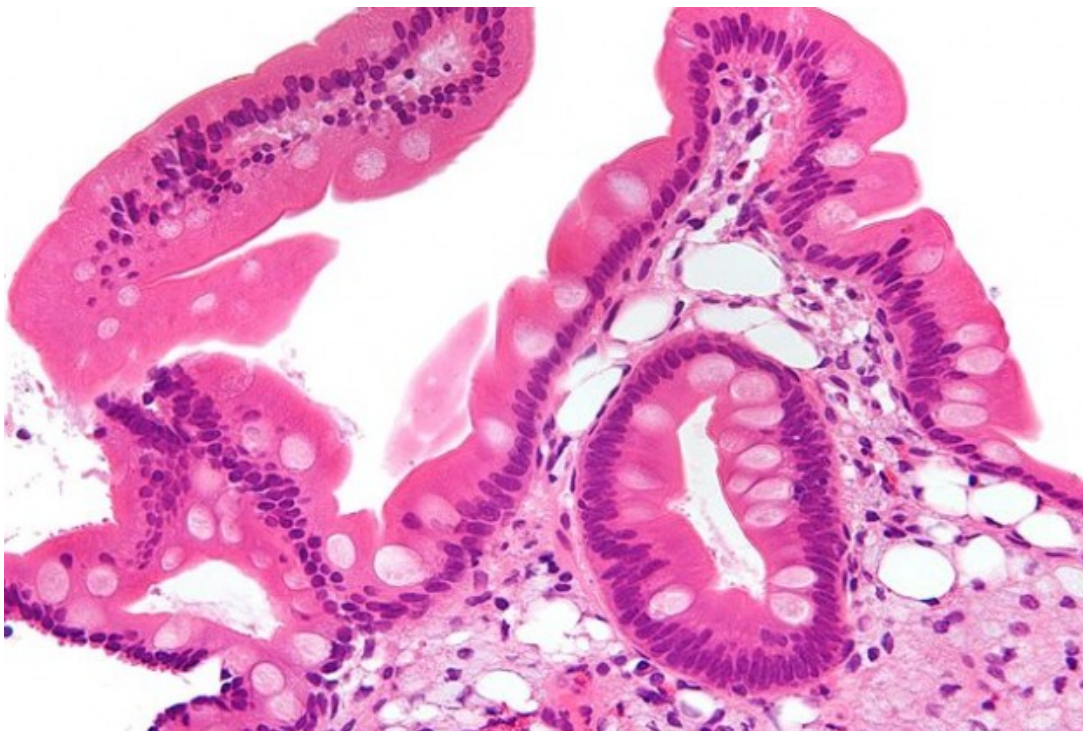
## Symptoms of Whipple's Disease

Early symptoms are **joint pain**. This symptom usually starts years before GI symptoms. As the disease progresses **malabsorption, diarrhea, and wasting occur**. Steatorrhea is also common. There are a variety of extra GI symptoms that occur in the later stages of the disease. These include **oculomasticatory myorhythmia** (repetitive movements of muscles in the face and eyes) uveitis, culture-negative [endocarditis](#), and hyperpigmentation of the skin. Dementia and memory loss are also common.

## Diagnosis of Whipple's Disease

Diagnosis is made by a **careful review of history and by biopsy**, which is definitive. Tissue samples from the duodenum will show foamy macrophages in the lamina propria that contain non-acid fast, gram-positive bacteria that are positive to ***Tropheryma whippelii*** antibody staining. **PCR** is a confirmatory test and can be used on synovial fluid, CSF, and blood samples. Additionally, mesenteric lymph nodes may appear enlarged on **abdominal CT**.

Small bowel biopsy showing PAS-positive granules in macrophages.



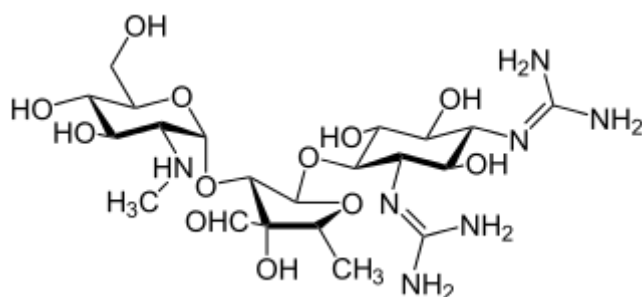
[Image](#): "Very high magnification micrograph of Whipple's disease. H&E stain. Duodenal biopsy. The images show the characteristic feature of Whipple's disease; foamy macrophages are present in the lamina propria." by Nephron. License: [CC BY-SA 3.0](#)

## Differential Diagnoses of Whipple's Disease

- Culture positive endocarditis
- Systemic lupus erythematosus
- [HIV](#) enteropathy
- [Celiac disease](#)
- Tropical sprue

## Therapy of Whipple's Disease

The treatment involves **antibiotic therapy** for up to one year. First, the patient is given streptomycin and penicillin for 14 days followed by co-trimoxazole (trimethoprim and sulfamethoxazole) for one to two years.



[Image](#): "Structure of streptomycin" by NEUROtiker. License: Public Domain

# Progression and Prognosis of Whipple's Disease

The chances of relapse after combined antibiotic therapy for one year is approximately 40 % and is more common with central nervous system symptoms. **Without treatment, mortality approaches 100 % after one year.**

## References

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