

Understanding HES

If you have received a diagnosis of hypereosinophilic syndrome (HES), you probably have many questions. This sheet can help you learn more about HES so you can take an active role in your treatment. Remember that you have reason to feel hopeful. Doctors learn more about HES every day.

What is HES?

HES is a group of diseases in which a high number of eosinophils are found in the blood. Eosinophils are a type of white blood cell that helps the body fight allergies and infections caused by parasites such as worms. Too many eosinophils can cause damage to the body's organs. Any organ can be affected, but the organs that are most often harmed are the heart, skin, and nervous system.

HES occurs more often in men than in women. It is most common in people aged 20 to 50 years.

How is HES diagnosed?

HES is diagnosed if:

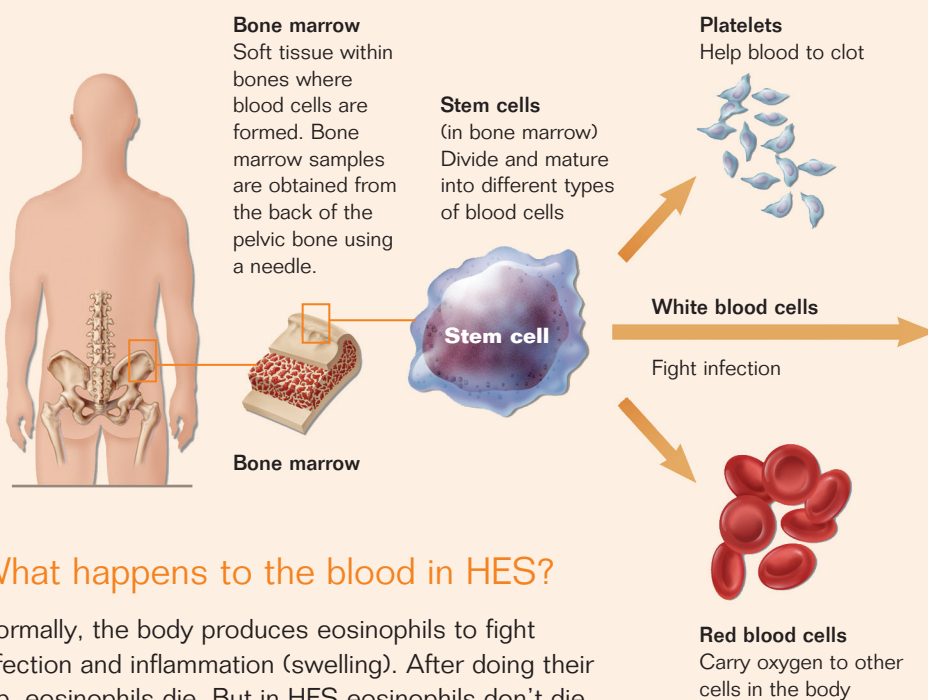
- ▶ Blood tests show a high number of eosinophils in the blood for at least 6 months
- ▶ There are signs of damage to the body's organs from having a high number of eosinophils for an extended time
- ▶ Other normal causes of high eosinophil levels, such as allergies or parasites, have been ruled out

Other tests may also be necessary, including:

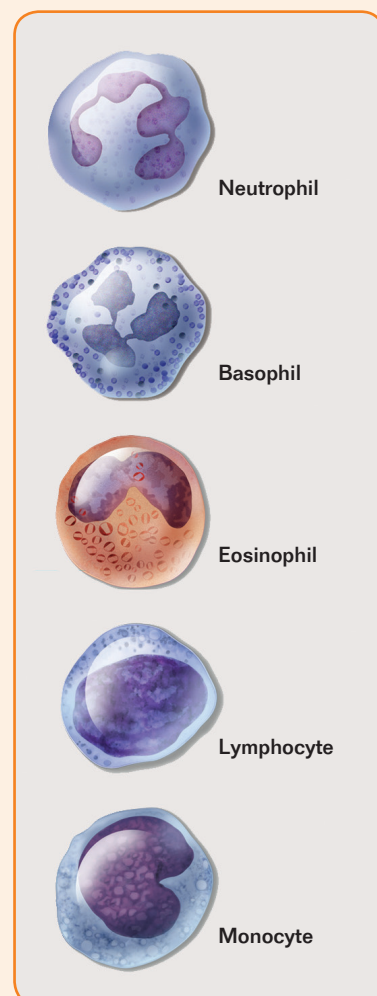
- ▶ **Heart tests**, such as an electrocardiogram and echocardiogram
- ▶ **Blood tests**, to check liver and kidney function
- ▶ **Skin tests**, called *biopsies*, in which a small piece of skin is removed from the body and examined under a microscope
- ▶ **Chest x-ray** of the lungs

How is blood produced?

Blood cell production begins in the bone marrow, where stem cells develop into different types of blood cells. Normal blood contains the right balance of red blood cells, white blood cells, and platelets to meet the body's needs.



Types of white blood cells



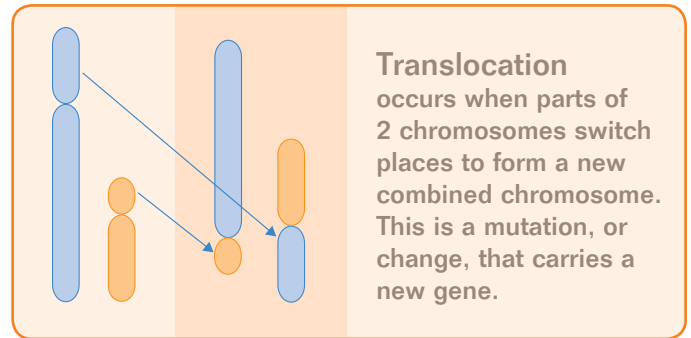
What happens to the blood in HES?

Normally, the body produces eosinophils to fight infection and inflammation (swelling). After doing their job, eosinophils die. But in HES eosinophils don't die when they are supposed to. The number of eosinophils in the blood rises. They build up in the organs. The organs become inflamed and are damaged.

What causes HES?

The cause of HES is not yet known. Research shows that in the myeloproliferative form of the disease (**myelo** means “bone marrow,” **proliferative** means “too much growth”), HES may be caused by a change in the genetic material on a chromosome. Chromosomes are found inside each cell of the body. They contain genes that tell each cell what to do in the body.

In HES, parts of 2 chromosomes switch places. This switch is called *translocation*. In other cases, a part of one chromosome is lost. This loss is called *deletion*. These changes result in the joining of 2 genes that were once apart. This combined gene makes a *platelet-derived growth factor receptor (PDGFR)*, which is a *tyrosine kinase*.



Tyrosine kinases are proteins in the body that cause cells to become active and to increase in number. In HES, abnormal tyrosine kinase activity stops eosinophils from dying when they are supposed to. Instead of dying, they build up and harm the body's organs.

How is HES treated?

Treatment for HES depends upon the form of the disease, how severe it is, and what organs have been affected. Some people with very mild disease whose hearts have not been affected may be closely monitored without treatment.

The goals of treatment are to:

- ▶ Decrease the number of eosinophils in the blood
- ▶ Prevent organ damage
- ▶ Slow down the disease

You and your doctor will choose the treatment that is best for you.

Treatment	How it works
Molecularly targeted therapy	Medicine treats the myeloproliferative form of HES Targets the source of HES by blocking the overactivity of abnormal tyrosine kinase, which causes eosinophils to build up in some cases
Glucocorticoids	Medicines treat the nonmyeloproliferative form of HES by reducing inflammation
Chemotherapy	Drugs kill abnormal cells or stop them from dividing Used when there is no response to glucocorticoids
Immunomodulators	Drugs help regulate the immune system and lower the number of eosinophils

Resources for learning more about HES

■ **American Partnership for Eosinophilic Disorders (APFED)**
USA
1-713-493-7749
www.apfed.org

■ **European Organisation for Rare Diseases (EURORDIS)**
France
33-1-56-53-52-10
www.eurordis.org

■ **National Organization for Rare Disorders (NORD)**
USA
1-203-744-0100
1-800-999-6673 (Voicemail only)
www.rarediseases.org

■ **Chronic Myeloproliferative Disorders (CMPD) Education Foundation**
USA
www.mpdinfo.org

■ **Leukaemia & Lymphoma Research**
UK
44-020-7405-0101
www.beatbloodcancers.org

■ **ORPHANET**
www.orpha.net