

Primary hemophagocytic lymphohistiocytosis (HLH)

# Understanding

## THE DIAGNOSIS

Finding out that you or your child have primary HLH can be overwhelming. Knowing some basic information about the condition can help you feel more in control as you prepare for treatment. This guide explains what primary HLH is and what happens inside the body. Don't hesitate to ask your doctor any questions about what you read.



### What Is GAMIFANT?

GAMIFANT is a prescription medicine used for the treatment of adults and children (newborn and older) with primary hemophagocytic lymphohistiocytosis (HLH) whose disease has come back or progressed, or other medicines have not worked well enough or cannot be tolerated.

### IMPORTANT SAFETY INFORMATION

**GAMIFANT can cause serious side effects, including infections.** GAMIFANT is a medicine that affects your immune system and may lower the ability of your immune system to fight infections. GAMIFANT may increase your risk of serious infections that can lead to death. These infections include tuberculosis (TB), histoplasmosis, herpes zoster infection (shingles), and other infections caused by viruses, fungi, or bacteria that can spread throughout the body. Your healthcare provider will:

- Test you for TB before you start treatment with GAMIFANT
- Treat you with a medicine for TB if you are at risk for TB or if you have a known positive TB test. Infections are common in people treated with GAMIFANT

**Please see Important Safety Information on pages 2-3. [Click here](#) for Medication Guide for Gamifant.**



## What Is GAMIFANT?

GAMIFANT is a prescription medicine used for the treatment of adults and children (newborn and older) with primary hemophagocytic lymphohistiocytosis (HLH) whose disease has come back or progressed, or other medicines have not worked well enough or cannot be tolerated.

## IMPORTANT SAFETY INFORMATION

### **GAMIFANT can cause serious side effects, including infections.**

GAMIFANT is a medicine that affects your immune system and may lower the ability of your immune system to fight infections. GAMIFANT may increase your risk of serious infections that can lead to death. These infections include tuberculosis (TB), histoplasmosis, herpes zoster infection (shingles), and other infections caused by viruses, fungi, or bacteria that can spread throughout the body. Your healthcare provider will:

- Test you for TB before you start treatment with GAMIFANT
- Treat you with a medicine for TB if you are at risk for TB or if you have a known positive TB test. Infections are common in people treated with GAMIFANT

### **Before starting GAMIFANT, tell your healthcare provider all of your medical conditions and if you:**

- Had TB in the past, or if you or a member of your family have been in recent close contact with someone with TB
- Have had a positive TB skin test (purified protein derivative test)
- Currently have an infection or have a history of infections, including histoplasmosis or herpes zoster (shingles)
- Are being treated for an active infection
- Have symptoms of an infection such as fever; sweat and chills; cough; breathing problems; blood in mucus (phlegm); or warm, red painful skin or sores on your body
- Are pregnant or plan to become pregnant. It is not known if GAMIFANT can harm your unborn baby
- Are breastfeeding or plan to breastfeed. It is not known if GAMIFANT passes into your breast milk. Talk to your healthcare provider about the best way to feed your baby during treatment with GAMIFANT

**Please see additional Important Safety Information on page 3.**

**[Click here](#) for Medication Guide for Gamifant.**



## IMPORTANT SAFETY INFORMATION (continued)

**Your healthcare provider will give you medicine to help prevent certain infections before you receive GAMIFANT.**

**Tell your healthcare provider about all the medicines you take,** including prescription and over-the-counter medicines, vitamins, and herbal supplements.

**After starting GAMIFANT,** tell your healthcare provider if:

- New symptoms of an infection appear
- Symptoms of an infection that you already had when starting GAMIFANT worsen. Your healthcare provider will monitor you closely for signs and symptoms of infections during treatment with GAMIFANT

### **GAMIFANT can cause serious infusion reactions.**

These are common, can also be severe, and can happen during or shortly after treatment with GAMIFANT. Your healthcare provider may temporarily stop your infusion and treat your symptoms before continuing your infusion if you have severe infusion reactions. Tell your healthcare provider right away if you get any of the following symptoms:

- Skin redness
- Chills
- Itching
- Chest pain
- Fever
- Shortness of breath
- Rash
- Nausea or vomiting
- Excessive sweating
- Lightheadedness or dizziness

**The most common side effects of GAMIFANT include** high blood pressure (hypertension) and fever.

**These are not all the possible side effects for GAMIFANT. For more information, ask your healthcare provider. You can also see the full Prescribing Information for GAMIFANT at <https://gamifant.com/pdf/Full-Prescribing-Information.pdf>.**

**To report suspected adverse reactions, contact Sobi North America at 1-866-773-5274 or FDA at 1-800-FDA-1088.**

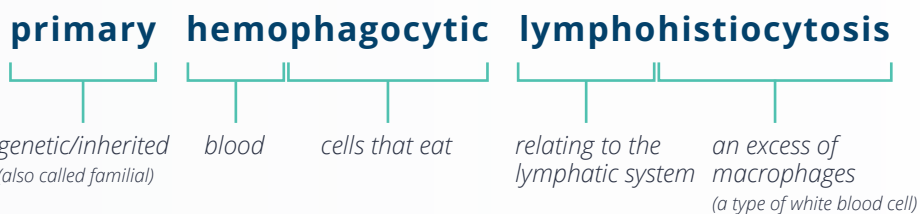
**Please see additional Important Safety Information on page 2. [Click here](#) for Medication Guide for Gamifant.**

## What is primary HLH?

Primary HLH is a rare genetic condition, which means it can be passed down in families. It usually affects infants and young children, but it can also affect older children and adults. Primary HLH is caused by a genetic mutation, or a change in the DNA. This mutation keeps the immune system from working properly.

- When the immune system works properly, it finds and attacks foreign invaders like viruses and bacteria to protect the body
- In people with primary HLH, the immune system doesn't work properly. It attacks the person's own cells and organs instead

### What does primary HLH stand for?



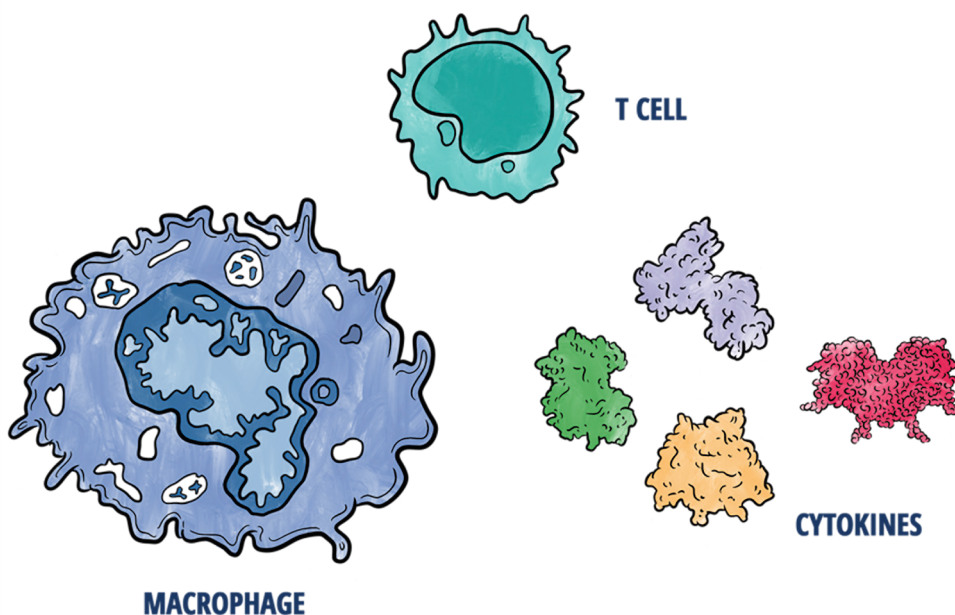
Watch a video on how primary  
HLH affects the body by visiting  
[GamifantCares.com/PHLH](https://GamifantCares.com/PHLH)

Please see Important Safety Information on pages 2-3.  
[Click here](#) for Medication Guide for Gamifant.

## What happens inside the body?

Everyone has immune cells. These cells travel throughout the body looking for harmful cells. When they find something suspicious, they bring it to the attention of T cells.

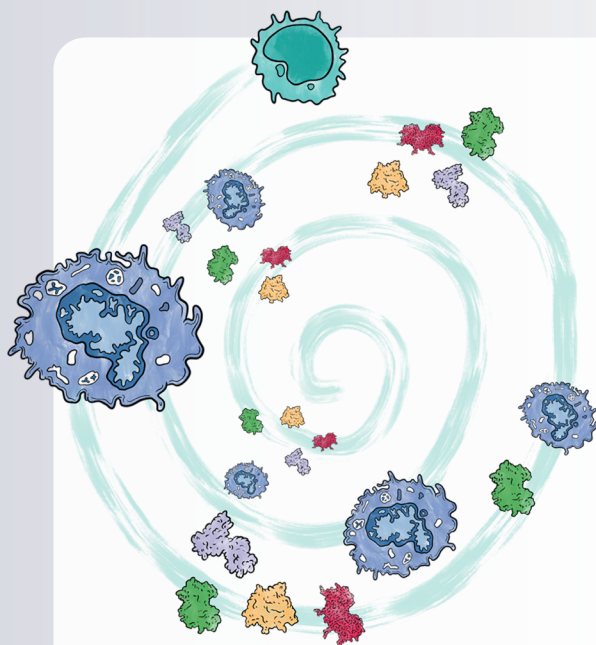
- T cells search out and destroy the harmful cells, but they need help. T cells release inflammatory messenger proteins called cytokines that activate macrophages to help the T cells get rid of the problem
- Macrophages are large cells that eat problem cells and white blood substances in the immune system. Macrophages secrete cytokines to help them with their job



Please see Important Safety Information on pages 2-3.  
[Click here](#) for Medication Guide for Gamifant.

## How does primary HLH cause inflammation?

T cells and macrophages usually work together to keep the body safe. But in primary HLH, they aren't able to destroy harmful cells. The body keeps trying by creating more macrophages and cytokines, which causes hyperinflammation.



- T cells keep releasing more and more cytokines, especially one called interferon gamma (IFN $\gamma$ )
- This activates more macrophages, and these macrophages release even more inflammatory cytokines
- The high level of cytokines creates a cytokine storm, which causes inflammation to get out of control
- This causes long-lasting fever, organ damage, and other symptoms of primary HLH

**Please see Important Safety Information on pages 2-3.**  
**[Click here](#) for Medication Guide for Gamifant.**

## What are the symptoms of primary HLH?

Primary HLH can be very serious. Being proactive and speaking with your doctor about any symptoms you or your child experience is very important.

**These are some of the signs and symptoms you and your doctor can look for or may have already noticed:**

- High fever that lasts a long time
- Rash
- Seizures or neck stiffness
- Yellowing of the eyes and skin (jaundice)
- Swollen lymph nodes (small round structures like your tonsils) that are part of the body's immune system and help fight infection or disease
- Low red blood cell or hemoglobin count (anemia) and looking very pale
- Coughing or trouble breathing
- Stomachache, vomiting, or diarrhea

*There may be other signs and symptoms of primary HLH not listed here.*

**If you think that your or your child's symptoms may be getting worse, tell your doctor right away.**

**Please see Important Safety Information on pages 2-3.  
[Click here](#) for Medication Guide for Gamifant.**



## Understanding the diagnosis is an important part of the treatment journey

We hope this information helps you feel more comfortable as you prepare for treatment.

*Learn more*

Visit [GamifantCares.com](https://GamifantCares.com) or call Gamifant Cares Support Services at 1.833.597.6530

**gamifant cares™**  
Support for the journey ahead



Ask your doctor for information about Gamifant as a treatment option and visit [GamifantCares.com](https://GamifantCares.com) to learn more.



Please see Important Safety Information on pages 2-3.  
[Click here](#) for Medication Guide for Gamifant.



Gamifant is a registered trademark, owned by Sobi AG and marketed by Sobi, Inc.  
©2022 Swedish Orphan Biovitrum. All rights reserved. PP-14786 04/22