talking with your doctor

ABOUT PRIMARY HLH

A diagnosis of primary HLH can be overwhelming and confusing. Primary HLH is not an easy disease to understand, and you probably have many questions. Your doctor is your partner in treatment and also the best source of answers to your questions.

THIS GUIDE CAN HELP YOU:

1 Understand some basic information about the disease

2 Organize your thoughts and questions

3 Get the most out of discussions with your doctor

HLH=hemophagocytic lymphohistiocytosis.

about primary HLH

"Primary hemophagocytic lymphohistiocytosis" is quite a mouthful to say, but don't worry, it's perfectly alright to call it "primary HLH."

Knowing some basic information about primary HLH can help you feel less confused and more in control. It may also help you feel more comfortable speaking with your doctor and asking questions.

The excess release of cytokines, including one called interferon gamma, leads to something called the cytokine storm, which causes inflammation to get out of control.

WHAT IS PRIMARY HLH?

It is a rare genetic disease that generally affects infants and children, although it can affect adults as well. In primary HLH, the immune system, which normally defends the body against foreign invaders, attacks the person's own cells and organs. Sometimes the disease can be controlled by medication alone, but bone marrow transplantation is the only known cure for primary HLH.

WHAT DOES "PRIMARY HLH" STAND FOR?

Primary hemophagocytic lymphohistiocytosis. Here's what it means when we break it down:

PRIMARY means that the disease could have been inherited from a parent to a child, which is why you may hear it referred to as "familial" HLH, though not all genetic errors that may cause it are known.

HEMO stands for blood, and **phagocytic** refers to phagocytes, which are cells that defend against foreign invaders.

LYMPHO means that it is related to the lymphatic system. The lymphatic system is made up of tissues and organs that produce cells to fight infections and other invaders.

HISTIOCYTOSIS means too many macrophages, which are a type of phagocyte (the defensive cells mentioned above). In primary HLH—since the body is being attacked by its own immune system—macrophages aren't able to fix the problem, but they continue to be activated and trigger the release of inflammatory cytokines. Cytokines are messenger proteins that communicate with other cells to trigger the body's response to pain or illness. In primary HLH, these signaling cells do not shut off, which leads to continuous inflammation throughout the body.

preparing for a discussion with your doctor

Since you or someone in your family have been diagnosed with primary HLH, you've probably had many thoughts and questions running through your mind. During your primary HLH journey, there may be various points when you'll feel the need to have a discussion with your doctor.

HERE'S WHEN 3 OF THEM ARE LIKELY TO OCCUR:

1
Shortly after diagnosis

2
During treatment

3
When you're ready to think about transplant

To get the most out of a discussion with your doctor, it can be helpful to write down your questions ahead of time. On the next page, you'll see some sample questions you might want to ask at various points in your journey. After each group of questions, there is space to list any additional ones you may think of and take notes during your discussions.

SOMETHING TO CONSIDER

When you're going to have a discussion with your doctor, it's a good idea to bring someone else along. That way, you'll have another set of ears to take in the answers and help you take notes. This is especially important when you're feeling nervous or emotional.

sobi
rare strength

questions you may have shortly after diagnosis

- Is there a cure for primary HLH?
- What did the different lab and imaging tests tell you?
- Which of the signs and symptoms my loved one or I are experiencing are due to primary HLH?
- Could other people in my family have the disease? Should we all get genetic testing?
- Is there a psychologist or social worker who could help us cope and manage this?
- What are the visitation rules at the hospital?
- What can I bring to the hospital to make my family member or myself more comfortable? Is there anything I shouldn't bring?
- Is there a community support group you recommend I get in touch with?
- Now that we have a diagnosis, what are our next steps?
- Who should I call when I have questions?

Add your additional questions in the space to the left.

questions you may have during treatment

- What kind of treatments will you use?
- How will we know if treatment is working and how often will you run tests to check?
- What are the potential side effects of various treatments?
- How long does treatment for primary HLH usually take?
- Will my loved one or I need to take medication for life?

Add your additional questions in the space to the left.

There's a short video about primary HLH called **"Max the Angry Macrophage." You can find it at bitly.ws/9bNg.**

sobi
rare strength

questions you may have about transplant

- What is a bone marrow transplant?
- How does a donor match work? Does the donor need to be an immediate family member?
- When will we find out if there's going to be a bone marrow transplant? What do you consider when you're deciding that?
- Is there another doctor or team of doctors we will need to work with?
- Will the transplant take place in this hospital or somewhere else?
- What kind of special care will be needed when my loved one or I return home after surgery?

Add your additional questions in the space to the left.

sobi
rare strength

your questions about primary HLH are important

A diagnosis of primary HLH can be scary and confusing, but you are not alone. Your doctor is your partner in treatment and can help you understand what to expect along the way.