talking with the doctor

ABOUT PRIMARY HLH

Finding out that your child has primary HLH can be overwhelming and confusing. Primary HLH is not an easy disease to understand, and you probably have many questions. Your child's doctor is your partner in treatment and also the best source of answers to your questions.

THIS GUIDE CAN HELP YOU:

- Understand some basic information about the disease
- Organize your thoughts and questions
- Get the most out of discussions with your doctor

HLH=hemophagocytic lymphohistiocytosis.

about primary HLH

"Primary hemophagocytic lymphohistiocytosis" is quite a mouthful to say. 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 equipped. It may also help you feel more comfortable speaking with your doctor and asking questions.

WHAT IS PRIMARY HLH?

It is a rare genetic disease that usually affects infants and children, although it can affect adults, too. Normally, the immune system defends the body against foreign invaders like viruses and bacteria. But in primary HLH, the immune system attacks the body's own cells and organs. Sometimes the disease can be controlled by medicine alone, but bone marrow transplantation is the only cure for most patients who have 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.

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

LYMPHO means related to the lymphatic system. The lymphatic system produces cells to fight infections.

HISTIOCYTOSIS means too many macrophages. Macrophages are a type of of phagocyte (defensive cells). In primary HLH, the body is being attacked by its own immune system. Macrophages aren't able to fix the problem like they normally would, but they keep trying to. This triggers the release of inflammatory cytokines.

Cytokines are messenger proteins that communicate with other cells. They trigger the body's response to pain or illness. In primary HLH, these cells don't stop sending messages. This leads to continuous inflammation throughout the body.

getting ready to talk with the doctor

Since your child has been diagnosed with primary HLH, you've probably had many thoughts and questions running through your mind. During the primary HLH journey, you'll have many conversations with the doctor.

HERE ARE 3 TIMES YOU SHOULD SPEAK WITH THE DOCTOR

Contact the doctor anytime a question or concern arises.

1
Shortly after diagnosis

2
During treatment

3
When you're ready to think about transplant

Don't hesitate to reach out to the doctor any time you have questions or concerns. To get the most out of your discussion, 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 others you may think of.

KEEP IN MIND

It's a good idea to bring someone else along when you're going to have a discussion with the doctor. 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. If your child is old enough, you may also decide to involve him or her in these discussions.

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questions you may have shortly after diagnosis

Use the spaces next to each question to take notes. Feel free to add any other questions you have.

- Is there a cure for primary HLH?
- What did the different lab and imaging tests tell you?
- Which signs and symptoms are due to primary HLH?
- Could other people in my family have the disease? Should we all get genetic testing?
- Is there a therapist 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 child more comfortable? Is there anything I shouldn't bring?
- Is there a community support group you suggest I get in touch with?
- Now that we have a diagnosis, what are our next steps?
- Who should I call when I have questions?

questions you may have during treatment

Use the spaces next to each question to take notes. Feel free to add any other questions you have.

- What kind of treatments will you use?
- How will we know if the treatment is working and how often will you run tests to check?
- What are the possible side effects of the treatments?
- How long does treatment for primary HLH usually take?
- Will my child need to take medicine for life?

There's a short video about primary HLH called **"Max the Angry Macrophage." You can find it by scanning the QR code below.**

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questions you may have about transplant

Use the spaces next to each question to take notes. Feel free to add any other questions you have.

- 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 child returns home after the transplant?

your questions about primary HLH are important

A diagnosis of primary HLH can be scary and confusing, but you are not alone. The doctor can help you understand what to expect along the way. Don't hesitate to ask any questions you have.

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