

CAREGIVERS

The Health Care Professionals You May Meet

A guide to who your loved one may meet along their journey with primary HLH

Many types of health care professionals are involved in the diagnosis and treatment of primary HLH. This guide serves as a resource to help you understand what each person does and how they may help your loved one.

HLH=hemophagocytic lymphohistiocytosis



Critical Care Specialist

About critical care specialists and why you might meet them

WHO ARE THEY?

Critical care specialists are doctors who diagnose, treat, and take care of people who are seriously ill or injured. They may treat a lot of different health issues and usually work in the intensive care unit of a hospital.

WHY MIGHT WE SEE THEM?

Primary HLH is a serious condition. Symptoms, such as inflammation and organ failure, can get worse very quickly and need to be taken care of right away. A critical care specialist may help treat symptoms and keep people stable while other doctors try to find what is wrong.

Your loved one may have seen a critical care specialist before you even knew the problem was primary HLH.

HOW LONG WILL WE SEE THEM?

If your loved one has already been diagnosed with primary HLH AND has moved out of the ICU, you may not see a critical care specialist again, unless symptoms get worse or organ failure happens.

WHAT WILL THEY DO?

- Provide emergency medical care to calm symptoms
- Perform life-saving procedures if necessary
- Give medicine to help with pain
- Keep a close watch on the patient to see how they are doing
- Share information with other doctors who will care for the patient

WHAT QUESTIONS SHOULD I ASK THEM?

- What happens next?
- What can we do to treat inflammation?
- What symptoms are you looking for?
- Is there anything we can do to make sure symptoms stay calm?
- What would cause my loved one to be in critical care again?

Hematologist/Oncologist

About hematologists/oncologists and why you might meet them



WHO ARE THEY?

Hematologists/oncologists are doctors who diagnose and treat blood diseases and cancers. They have received special training in the study of blood and cancer and may have training in bone marrow transplants.

WHY MIGHT WE SEE THEM?

Your loved one may see a hematologist/oncologist to help figure out what is causing their symptoms. Because symptoms of primary HLH are very similar to some types of cancers or infections, a hematologist/oncologist can help rule out other conditions.

HOW LONG WILL WE SEE THEM?

Hematologists/oncologists specialize in primary HLH. So, if your loved one is diagnosed with primary HLH, this is the main doctor that will oversee treatment. They will also decide if your loved one needs a transplant and will provide the care for it – not everyone will need a transplant.

WHAT WILL THEY DO?

- Tests to rule out cancer or other conditions that may be causing symptoms
- Tests to see what may be making your loved one sick: Ferritin level testing (a protein that stores iron), bone marrow testing, blood and liver function testing

If they think your loved one has primary HLH:

- Check if they have 5 or more of the signs and symptoms
- Order genetic tests
- Start treatment (*because of the seriousness of primary HLH, they may do this while still waiting for test results*)

WHAT QUESTIONS SHOULD I ASK THEM?

- How did you diagnose primary HLH?
- Can you review the test results with us?
- Are you the main person who will be managing primary HLH for us? Who else will be involved?
- What are your thoughts about a bone marrow transplant? Can you explain that process?
- Will we need treatment before a bone marrow transplant?
- Will we continue seeing you for primary HLH?

Hepatologist/Gastroenterologist

About hepatologists/gastroenterologists and why you might meet them

WHO ARE THEY?

Hepatologists/gastroenterologists are doctors who diagnose and treat diseases of the digestive organs (liver, stomach, intestines, pancreas, and gallbladder). Hepatologists focus specifically on the pancreas, gallbladder, and liver.

WHY MIGHT WE SEE THEM?

Your loved one may see a hepatologist/gastroenterologist due to symptoms of primary HLH, such as:

- A swollen spleen and liver
- Poor liver function
- Yellow skin or eyes (jaundice)

The hepatologist/gastroenterologist can help rule out illnesses related to the digestive system.

HOW LONG WILL WE SEE THEM?

Once a hepatologist/gastroenterologist rules out digestive conditions, they are likely to send your loved one to another specialist to make the diagnosis. However, they may still check in during treatment since primary HLH can affect the spleen and liver.

WHAT WILL THEY DO?

- Ask questions about your loved one's medical history and the health of their family members
- Do a physical exam of the abdomen and digestive tract to find out where symptoms are coming from
- Use a small camera to look inside the body (this is called an endoscopy)

WHAT QUESTIONS SHOULD I ASK THEM?

- What is impaired liver function?
- What does an enlarged spleen mean?
- What is jaundice?
- What can be done to fix inflammation in the spleen/liver?
- What treatment do you think we need?
- What labs do you use to monitor my liver?

Infectious Disease Specialist

About infectious disease specialists and why you might meet them



WHO ARE THEY?

Infectious disease specialists are doctors who are experts in diseases caused by bacteria, viruses, fungi, and parasites. They help diagnose and treat serious infections and may work with other doctors to diagnose and treat other illnesses.

WHY MIGHT WE SEE THEM?

Certain symptoms of primary HLH may be similar to viral infections or sepsis (where organs can shut down because of an infection). The following symptoms of primary HLH may be confused with infectious disease. The infectious disease specialist will work with other doctors to rule out infections and other conditions that cause these symptoms, like:

- High fever that lasts a long time
- Seizure or neck stiffness
- Swollen lymph nodes
- Coughing and difficulty breathing
- Rash
- Confusion
- Enlarged spleen
- Vomiting or diarrhea

HOW LONG WILL WE SEE THEM?

Your loved one may continue to see an infectious disease specialist once they are diagnosed with primary HLH. This is may be able to help manage symptoms. Also, people with primary HLH have weaker immune systems, which means they may be more likely to get an infection. The infectious disease specialist can help treat or prevent infections.

WHAT WILL THEY DO?

- Review symptoms and physical exam results
- Take blood, urine, or other fluid samples to check for an infection
- Refer the patient to the right specialist if all infectious diseases are ruled out

WHAT QUESTIONS SHOULD I ASK THEM?

- Do you think my loved one has an infection?
- How will we treat these symptoms?
- Do they need antibiotics? Which ones and for how long?
- What are the side effects of the medication you are prescribing?
- What other treatments do you suggest?
- Will we continue to see you for primary HLH?
- How can we prevent any future infections?

Rheumatologist

About rheumatologists and why you might meet them

WHO ARE THEY?

Rheumatologists are doctors who diagnose and treat conditions that affect muscles, bones, and joints. They also treat autoimmune diseases, which are diseases where the immune system attacks a person's own cells and organs.

WHY MIGHT WE SEE THEM?

In primary HLH, the immune system does not work right, attacking the person's own cells and organs and causing inflammation.

The rheumatologist will help decide if inflammation is caused by primary HLH or other conditions that are similar.

HOW LONG WILL WE SEE THEM?

Your loved one may continue to see a rheumatologist after being diagnosed with primary HLH. They may take care of them or send them to a hematologist/oncologist. Sometimes the two types of doctors will work together.

WHAT WILL THEY DO?

- Ask questions about your loved one's medical history and the health of their family members
- Do a physical exam to find the cause of the problem
- Perform tests to rule out other illnesses that are similar to primary HLH

If they think your loved one has primary HLH:

- Check if they have 5 or more of the signs and symptoms
- Order genetic tests
- Ask a hematologist/oncologist for an opinion

WHAT QUESTIONS SHOULD I ASK THEM?

- How did you diagnose primary HLH?
- What happens next?
- Will we continue to see you?
- What treatment do you think we need?
- What can we do to help the inflammation?
- Should we also see a hematologist/oncologist?

Social Worker

About social workers and why you might meet them



WHO ARE THEY?

Social workers are trained professionals who often work in a hospital setting to help patients and their families understand a particular illness.

WHY MIGHT WE SEE THEM?

Symptoms of primary HLH can be serious. People are usually admitted to the hospital for further evaluation. Also, primary HLH can be difficult to diagnose and may be mistaken for other conditions. This can feel scary and overwhelming for patients and caregivers. A social worker can help you get the support you might need during this time.

HOW LONG WILL WE SEE THEM?

A social worker may help your loved one while they are admitted to the hospital and after they go home.

WHAT WILL THEY DO?

- Help you and your loved one adjust to hospital admission
- Help you and your loved one understand primary HLH and treatment options
- Tell you and your loved one more about different doctors and care team members you may meet
- Help you and your loved one communicate with each other and your health care team
- Help you and your loved one work through emotions and offer counseling for decisions
- Arrange resources/funding to help with affording treatment
- Coordinate patient discharge from the hospital

WHAT QUESTIONS SHOULD I ASK THEM?

- What resources are available to support us?
- Are there counseling services for me or my loved one?
- How can I find help paying for treatment?
- What's the best way to communicate with our care team?
- Do you recommend any support groups in the community?
- What should we do about treatment and care once we're not in the hospital?
- Will I still be able to communicate with you after leaving the hospital?

Your Questions About Primary HLH Are Important.

A diagnosis of primary HLH can be worrisome and confusing for your loved one, but they are not alone. The doctors can help you and your loved one understand what to expect along the way. Don't hesitate to ask any questions you may have.

