



The Dragons Inside

Understanding Hemophagocytic Lymphohistiocytosis (HLH)

A guide for you and your family

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Hello there!

We're guessing you've started reading this book for two reasons.

The first is because you like dragons. Who doesn't like dragons?! For a start, all that fire-breathing action is so cool!

They come in all shapes, sizes and colors. Red, green, purple, blue...

And that swooshing tail that can knock buses flying! Wouldn't it be great if we had our own pet dragons to zoom around on?! Imagine how quickly you would get to school!

The second reason is that someone in your family has been told they have **Hemophagocytic lymphohistiocytosis**. This could be your brother or sister, maybe your cousin.

We should point out right away that if you can't say Hemophagocytic lymphohistiocytosis, you're not alone. Looking at those two words puts your head in a spin, right? **That's why we shorten it to HLH.**

As luck would have it, dragons can help to explain everything you need to know about HLH.



Hello parents or carers

This guide to HLH is designed to help you explain the condition to your young children or family members. Throughout this guide you'll find extra information in boxes just like this one.

HLH is a rare immune disorder. There are two main types of HLH: primary (also called familial) and secondary. Primary HLH is caused by a genetic mutation (a faulty gene) that is inherited. It usually shows up within the first year of life. Secondary HLH is not inherited and develops as a result of another illness, such as an infection.



Your dragons inside

Ever wished you had your own dragons?!
Well, get ready for some good news...

You do!

Before you start worrying about whether your back yard is big enough to keep them in, you should know that these dragons are not flying about above us. They're not swooping down on castles or filling the sky with fire. **No, these dragons are far cooler than that.**



Dragons live inside us

They are tiny dragons. So miniscule that we can't see them without a super powerful magnifying glass. We all have them. Your mom has them, so does your dad. Your brothers, sisters and cousins have them too.

Now, we know what you're thinking:

What do we need dragons for?!?!

Let us explain.

Your body is like a castle

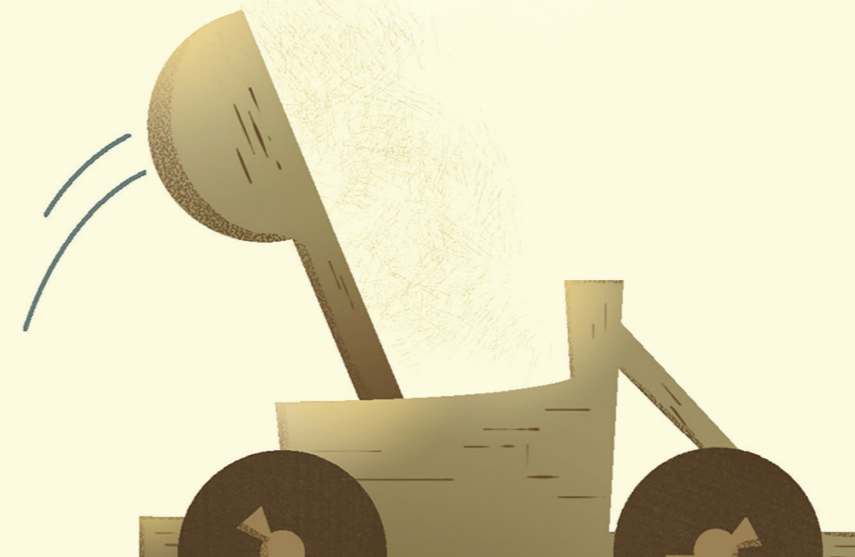
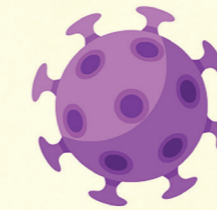
The huge stone walls are your skin. This helps to keep all the things you need inside, like your heart and lungs. You don't want to misplace those. Those walls also stop things from entering. Things like those nasty bugs that give us tummy aches or stinking colds!

What would your dragon look like?

Draw it here. Need inspiration?! Check out the dragons on the next page.

We know these bugs as bacteria and viruses. Sometimes those sneaky bugs do find a way past the walls and into the castle.

And this is why we need dragons. Lots of them.



Say hi to the dragons

There many types of dragon who help guard your castle.

Each dragon has its own special role. Here are the important ones to know.

Spy dragons

These are really two dragons!
The Small Spy Dragons keep a look out.
The Large Spy Dragons keep a record of all the viruses and bacteria seen in their Big Book of Bugs.



Ninja dragons

These are expert fighters.
No bug wants to fight these!

Ka-Pow!

Which is your favourite dragon?

Hungry dragons

These gobble up all the bacteria and viruses.

Munch! Nom! Nom!



Your army of dragons

These happy dragons form an army to keep your castle safe. They work together to hunt down the bugs and remove them. The last time you had a tummy ache, it was your dragons who helped you feel better. **This army of dragons are known as the immune system.**

What happens when a bug sneaks past the castle wall?



Spy Dragons are the first to spot them. When the **small Spy Dragon** spots a bacteria or virus it sounds the alarm. To do this it sends secret smoke signals to the other dragons.

The larger Spy Dragon uses his Big Book of Bugs to find out what type of bug has been seen.



Ninja Dragons arrive really quickly. These dragons are expert fighters! They breathe a lot of fire. They use their ninja skills and all that fire to capture the bugs and viruses.

The huge Hungry Dragons then show up. The only thing bigger than a Hungry dragon is its own appetite!

Hungry Dragons just love to munch on bacteria and viruses. They will snack on bugs all day long.

Together these dragons stop the bad bugs from damaging the castle!

Can you decode the small Spy Dragon's smoke signals??

A	B	C	D	E	F	G
H	I	J	K	L	M	N
O	P	Q	R	S	T	U
V	W	X	Y	Z		



ANSWER: COME QUICK



For parents and carers - Immune cells

These dragons represent just some of the **immune cells** in our bodies. There are many types of immune cell, each with a specific role to play when fighting infections.

Spy dragons are known as **T cells**, Ninja dragons as **NK cells**, and Hungry dragons as **Macrophages**. Collectively these are referred to as **lymphocytes** or **white blood cells**.

These are the three types of immune cells involved in HLH, but you may also have heard of other immune cells such as B cells, neutrophils, and monocytes. These are not involved in HLH.

Sometimes dragons get confused

Dragons are really hard workers. But sometimes children are born with dragons that get easily confused. This is very rare but does happen.

When Spy Dragons, Ninja Dragons and Hungry Dragons are confused they do things they shouldn't.

They breathe more fire and snort more smoke than normal.



They talk more and start asking other dragons to breathe more fire too!



Confused dragons can damage the castle

The extra fire, the swooshing tail and all that smoke can cause damage to the castle. When this happens, a child can start to feel unwell.

Doctors call this **hyper-inflammation**. It is these confused dragons (hyper-inflammation) that cause HLH.

They munch on the castle wall and bash it with their huge, swooshing tail!



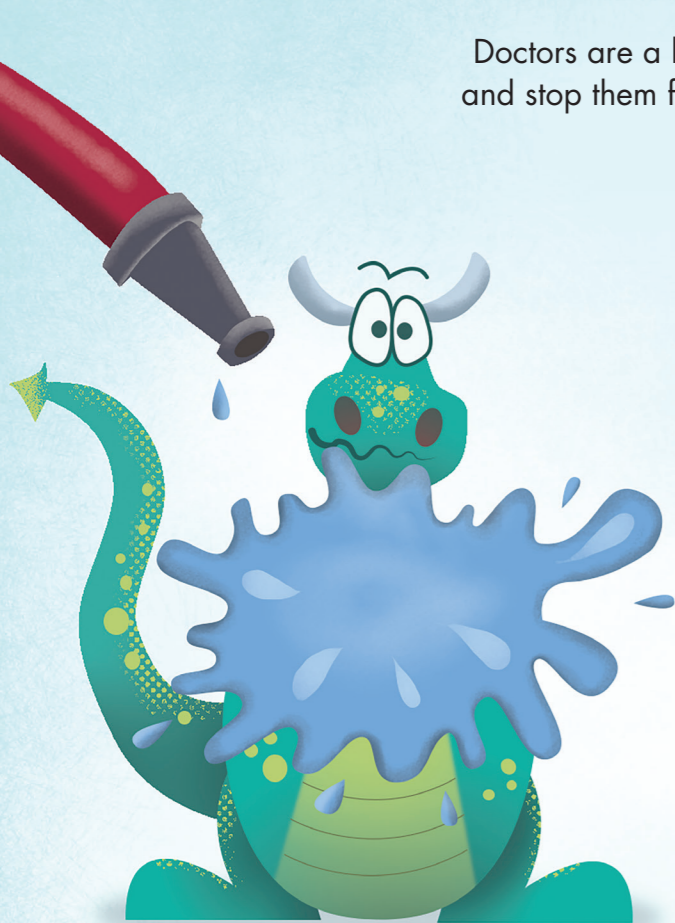
For parents and carers - Hyper-inflammation and HLH

The T cells, NK cells and macrophages in patients with HLH can't fight infections like they normally would. This means the immune system becomes poorly regulated, over stimulated and overactive. This leads to hyper-inflammation, where immune cells start to damage tissues and organs, such as bone marrow and the liver.

Calming dragons down again

Doctors are a bit like wizards. They have a number of medicines to calm dragons and stop them feeling so confused. Doing this can help a child with HLH feel better.

Some ways to calm a dragon



Put out the fire!

Stopping the dragon from breathing fire means it can't damage the castle anymore.



Make them sleepy!

A sleepy dragon can't snack on the wall or swoosh its tail against it.



Stop the chitter-chatter!

If the dragons can't talk, they can't ask other dragons to breathe more fire.

Can you help the dragon fall asleep?

Join the numbers to see what the dragon is doing.



For parents and carers – Treating HLH

Given the immune system is overactive in HLH, the main priority of treatment is to suppress or dampen down the immune system. This reduces the risk of tissue and organ damage. There are a number of different treatments available that can calm the immune system. Each works in a slightly different way, but all aim to suppress the hyper-inflammation and prevent further damage. Your child's doctor will explain about the different treatment options, how they are administered and how they work. These treatments usually put HLH into remission, but the risk of a relapse does remain.



Spotting confused dragons is really hard!

Doctors like to keep an eye on confused dragons. They also like to check to see if their medicines are working and calming the dragons.

But remember that we said dragons are really, really tiny? Well, this makes it hard to see them and know if they are confused or not.

So how do we know?!?

Doctors are experts at spotting confused dragons. Doctors don't actually see the dragons. Instead, they look for signs or signals that they are there.

To do this, doctors perform lots of tests. But these are nothing like your school tests!

These tests need to be done in a hospital.

This is the reason why you and your parents spend time there.

The tests can tell a doctor if there are:

Too many secret smoke signals

Too many dragons

Too much dragon fire

Damage to the castle



Can you help the doctor spot the confused dragons?

Hidden in the castle are **8** confused dragons. Can you find them?



For parents and carers – How is HLH monitored?

Blood samples are taken. These blood tests are used to monitor the blood cell levels and the health of the other organs, to see if the disease is responding to treatment or not. Blood tests also monitor for complications like new infections. Other samples, such as bone marrow or lymph node tissue, may also be taken. Imaging tests are also used.

Sharing dragons

Even with those medicines, sometimes a child with HLH can still feel poorly. But don't worry, your team of doctors and nurses still have another trick up their sleeve...

They can share dragons!

We're all different. You don't look exactly the same as your mom or dad or brother or sister. But you kinda do, right? You might have a little nose like your mom. Big ears like your dad. Scruffy brown hair like grandpa.

Dragons are like this. Everyone has different dragons. But some people have dragons that look similar. When this happens, doctors can remove the confused dragons. They then take calm dragons from one person, and give them to a child with HLH. This can help the child feel better.

Would you like to share a dragon?!

Then see the next page!



For parents and carers – Stem cell transplants

Once your child's hyperinflammation has been brought under control by their treatments, your doctor may recommend a hematopoietic stem cell transplant (HSCT), which is also known as a bone marrow transplant (BMT). This offers the chance of a long-term cure from HLH. HSCT aims to replace the faulty immune cells with healthy ones.

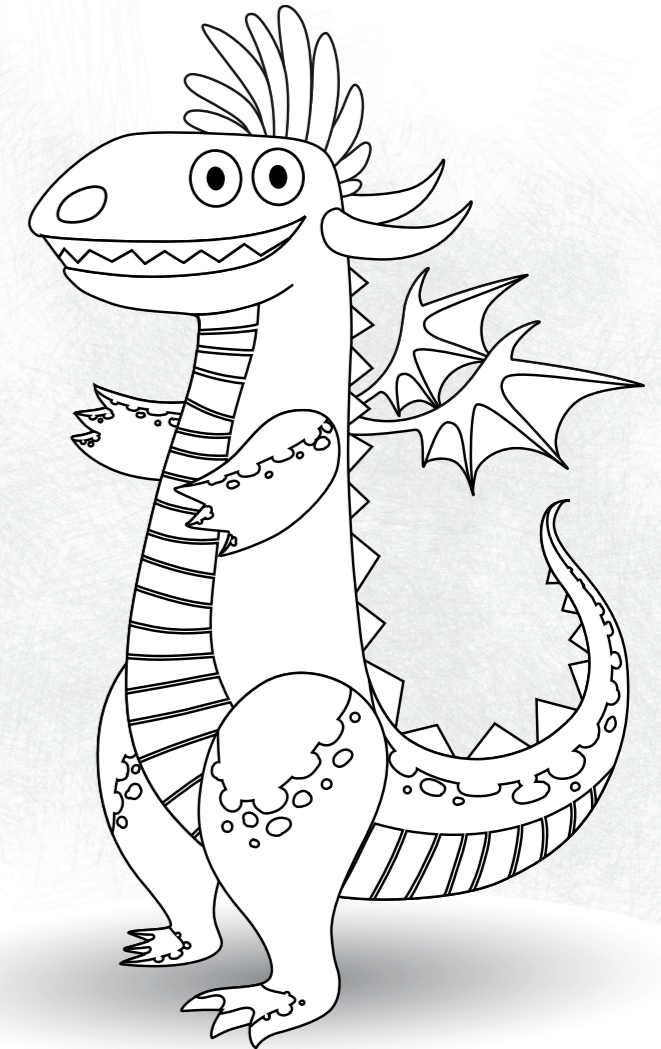
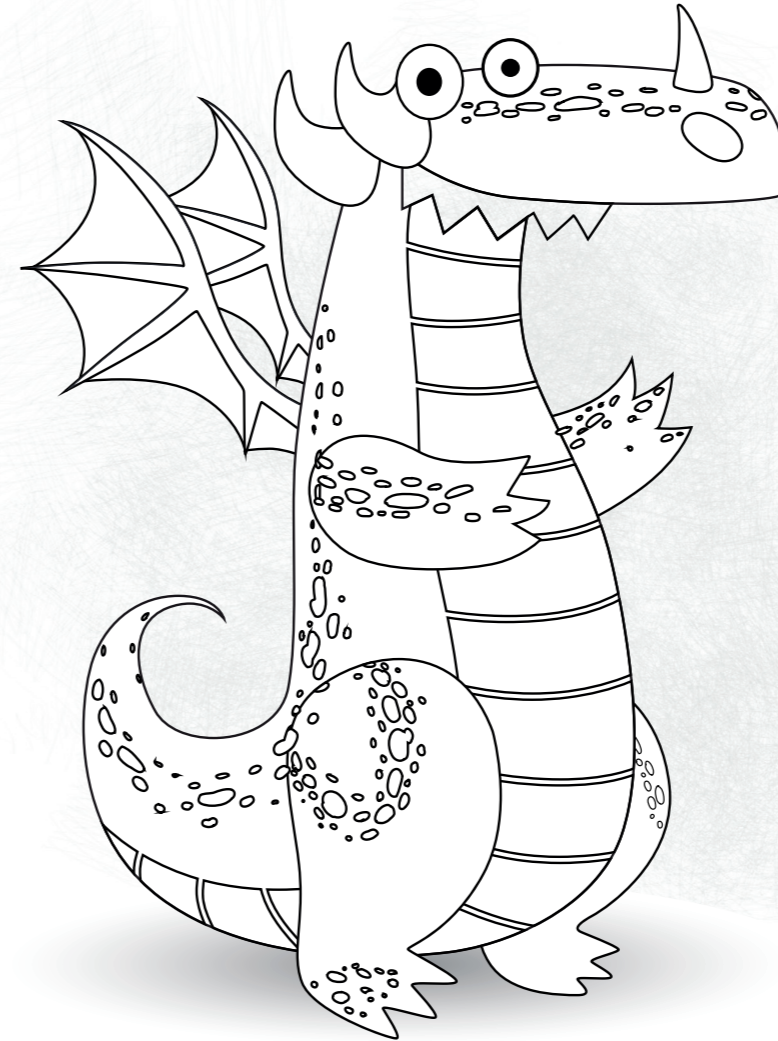
Hematopoietic stem cells are cells that can develop into any type of white or red blood cell. They can be obtained from the healthy donor. To prepare for the transplant, the child with HLH will be given medicines (known as conditioning). When they are ready for the transplant, the healthy stem cells from the donor are given by transfusion into a vein. After a few days or weeks, the stem cells develop into healthy blood cells, including healthy macrophages, NK cells and T cells.

A donor can be a sibling, close family member or an unrelated donor. Doctors will perform tests to see if someone is a close match for your child.

A HSCT may not be recommended for all patients with HLH. Your doctor will be able to explain if HSCT is an option for your child.

Why not share a dragon?

Below are two calm dragons. Color them both in, then cut them out and give one to your brother or sister.



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Dear

I thought you might like this picture
of a dragon I made just for you :)

From

Dear

I thought you might like this picture
of a dragon I made just for you :)

From

What does that word mean?!?

Biopsy: Surgical removal of a small piece of tissue. This is examined under a microscope to diagnose a disease.

Bone marrow: Soft, spongy tissue that is located in the centre of our bones. Bone marrow contains stem cells that develop into blood cells and all the cells of the immune system.

Bone marrow transplant (BMT): This involves the transfer of stem cells from bone marrow. The donor can be related or unrelated. The new bone marrow replaces the recipient bone marrow, giving the recipient a new immune system (see also Hematopoietic stem cell transplant).

Donor: A person who could donate bone marrow or stem cells to another person. Donors are often family members, but they can also be unrelated. A donor needs to be well matched with the potential recipient. Whether a person is a match or not is decided by tissue-typing.

DNA: This stands for Deoxyribonucleic Acid. DNA contains our genetic information (see Gene).

Gene: A section of DNA that codes for a protein. Genes are the units of inheritance that parents pass on to their children. Sometimes a piece of DNA can have a mistake (or mutation).

Hematopoietic stem cells: Cells that will develop into blood cells or immune cells.

Hematopoietic stem cell transplantation (HSCT): Hematopoietic means 'blood-forming'. This involves the transfer of bone marrow to a recipient. The donor cells are given by intravenous infusion and give the recipient a new immune system (see also Bone marrow transplant).

Immune system: The organs, tissues and cells (e.g., white blood cells such as macrophages, NK cells and T cells) that protect the body against infection and disease.

Inflammation: A natural immune response that helps defend the body against harm from invaders like bacteria and viruses. Sometimes the body can trigger inflammation where there is no invader.

Inheritance: The passing down of genetic information from parents to their children.

Intravenous infusion: Medicines that are given by infusion directly into a vein.

Macrophage: A type of white blood cell that is important for destroying invading bacteria and viruses.

Mutation: A change in a gene or group of genes. Many mutations cause no harm, but others can cause problems, such as primary HLH.

NK cells: Also known as Natural Killer cells. These are a type of white blood cell and are involved in killing viruses and bacteria.

Remission: When the signs and symptoms of a disease are controlled. This means a person no longer feels ill, but does not mean the disease has been cured.

Systemic hyper-inflammation: Severe inflammation throughout the body caused by the body's own immune system.

T cells (or T lymphocytes): A type of white blood cell. They are involved in the second phase of the immune system, and can have many functions: they can kill infected cells, co-ordinate other immune cells and regulate the immune response.

White blood cells: A group of small, colorless blood cells. There are many types including monocytes (e.g., macrophages), lymphocytes (e.g., T cells), neutrophils, eosinophils and basophils. They all play a major role in the immune system.



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With special thanks to

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For more information about these organizations please visit:

<https://www.aileonlus.org/>

<https://www.cincinnatichildrens.org/service/h/hlh>

<https://www.matthewandandrew.org/>

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