

# A GUIDE FOR CARERS OF CHILDREN LIVING WITH PTLD

This information guide is for parents and carers of children diagnosed with post-transplant lymphoproliferative disease (PTLD) following solid organ transplantation (SOT) or allogeneic hematopoietic cell transplantation (HCT), also called bone marrow or stem cell transplantation.

**This leaflet has been created in collaboration with the Lymphoma Coalition a worldwide network of patient groups.**

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## Disclaimer

*The information in this leaflet provides helpful information about PTLD. Family members may also find it beneficial to read.*

*This material does not replace the advice of a healthcare professional. Be sure to ask your healthcare professional any questions or concerns you have.*



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This guide is primarily aimed at the parents and carers of a child recently diagnosed with PTLD. By 'child', we mean someone between two to 18 years of age, and by 'carer', we mean anyone supporting a child with PTLD. We provide tips to help you with challenges and suggestions for further support and information. This guide can also help explain the condition to siblings, young relatives and friends of a child with PTLD.

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# YOUR CHILD IS DIAGNOSED WITH POST-TRANSPLANT LYMPHOPROLIFERATIVE DISEASE (PTLD)

It is unexpected and can be overwhelming when your child is diagnosed with post-transplant lymphoproliferative disease (PTLD) after their long transplant journey. Various emotions might come into play, such as fear, confusion, denial, and possibly anger. The challenges you face daily, supporting a child with PTLD will be more manageable with the right advice and information.

PTLD is a rare and potentially life-threatening complication of solid organ transplantation (SOT) or allogeneic hematopoietic cell transplantation (HCT)<sup>1</sup>. PTLD is well known<sup>2</sup>, and your child's medical care team will have a treatment plan in place. It can be treated successfully, especially if diagnosed early<sup>3</sup> as treatment options have improved dramatically in the last few decades<sup>2</sup>.

Increasing your awareness and understanding of PTLD will help you and your child manage your day-to-day life reduce stress, anxiety and related treatments.



# WHAT IS PTLD?

PTLD, or post-transplant lymphoproliferative disease, is the name of a group of disorders ranging from benign growths of affected tissue to lymphomas that sometimes develop following solid organ transplantation (SOT) or allogeneic hematopoietic cell transplantation (HCT), also called bone marrow or stem cell transplantation<sup>1</sup>.

When a child has been transplanted, doctors treat them with drugs to suppress their immune system. PTLD occurs because the immune system is weakened and, therefore, ineffective at controlling certain viruses, such as the Epstein-Barr virus (EBV)<sup>4</sup>. EBV is a common virus that doesn't usually cause problems in someone with a standard immune system, but in a weakened system, EBV can remain uncontrolled and lead to PTLD<sup>5</sup>.

### Symptoms of PTLD<sup>3,5</sup>

The symptoms of PTLD depend on where in the body the disease begins and how far it has spread when it is found. General symptoms might include:

- Fever
- Unexpected weight loss over a few weeks
- Swollen lymph nodes (glands)
- A general feeling of poor health
- Night sweats
- Fatigue

The most common is a painless lump, usually in the neck, armpit, or groin. This is a swollen lymph node (gland). Your child might have swollen lymph nodes deep within the body, where they can't be felt from the outside.



# HOW CAN PTLD BE TREATED?

The treatment your child needs depends on the type of PTLD and how widespread it is<sup>3</sup>. Often the first line of treatment is to reduce or completely stop the immunosuppression medicine causing the weakened immune system; however, that is not always possible with the child's clinical status. Restoring the immune system to its normal function can successfully treat PTLD<sup>4</sup>.

Other treatment approaches include<sup>4,6</sup>:

<b>Antibody therapy</b>	Antibody therapies help the immune system recognize and fight the target cells (e.g tumor cells, infected cells).
<b>Chemotherapy</b>	The chemotherapy treatment depends on the type of PTLD. Chemotherapy is often given with antibody therapy called chemo-immunotherapy.
<b>EBV-specific T-cell therapy</b>	For patient diagnosed with EBV+ PTLD, anti-EBV specific T cell.
<b>Clinical trials</b>	A clinical trial is a medical research study sometimes involving children and can be an important treatment option. Ask your child's medical team any questions about clinical trials or visit <a href="https://clinicaltrials.gov/">https://clinicaltrials.gov/</a> for more information on studies being conducted worldwide.

### Who provides the treatment plan?

Your child's treatment plan will be created by a multidisciplinary team (MDT) composed of:

- Transplant physicians
- Haemato-oncologists
- Pediatricians

They consider your child's general health status, PTLD status, the degree of immunosuppression, and the types of therapy available.

**Ask the medical care team if you have questions about your child's PTLD treatment plan.**



# HOW DO I CARE FOR MY CHILD?

Receiving a diagnosis that your child has PTLD can be challenging to process, especially after the trauma of the transplant and all that it entails. Most people who look after a child with PTLD don't know anything about it before diagnosis. There's so much to process, intellectually and emotionally. Different families have different ways of coping, but there are some sound strategies that anyone can employ.

## Key things to remember:

- Educate yourself about your child's condition
- Create a support network for emotional and practical support
- Be patient with the pace of treatment and recovery
- Do things that help you remain positive

## Shock and confusion

When a doctor tells you your child has PTLD, it's natural to lose focus on what else the doctor is saying and block out other, often complicated, information about your child's illness. The medical care team will understand, so don't be afraid to ask them questions and ask for support; you need to understand what PTLD is and what to expect. You might find it helpful to keep a notebook with important information.

## Stress and emotions

It's essential to manage your feelings and keep stress in check. You may feel sad, angry and overwhelmed, which is quite normal. You may feel a loss of control over making your child feel better or about the future prognosis. Addressing your child's condition directly is the best way to move forward. A study of parents of children with cancer found that those parents who took action and focused on the problem experienced lower levels of anxiety and depression than parents who denied or avoided the situation<sup>7</sup>.

## Enlist help

Build a support network you can rely on to help ease your load; relatives and friends can often be a source of strength and understanding. If you have other children, make them part of the team; help them figure out how they can care for their brother or sister. Tell extended family and friends as well. It's worth remembering that people will want to support you but can only help if they know what you're facing.

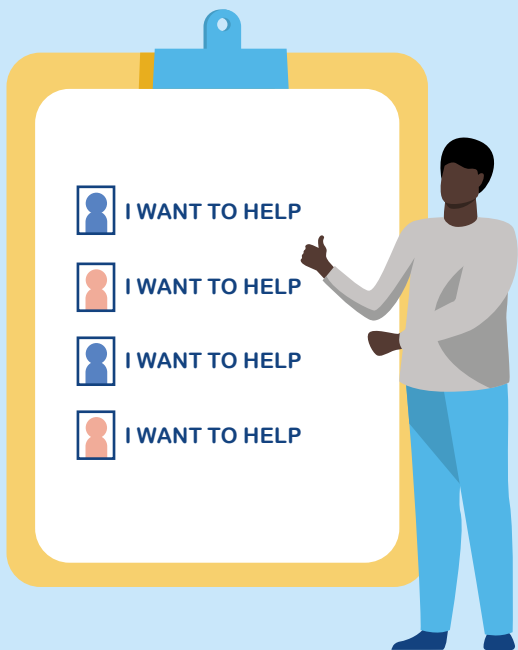
Hope plays an important role in the ability to cope. Talk with other parents whose children have had a similar diagnosis or treatment and recovered.

## The medical care team

Communicate regularly with your child's medical care team to get the necessary answers and clear doubts. Expressing your child's needs and preferences will help the care team make the best choices.

## Look after yourself

It's essential to look after yourself and stay physically and emotionally well. Maintain your own life, have your own space and take time to recover and rest. Don't feel guilty about taking some time for yourself. In the long run, doing so will help you and your child.



# HOW SHOULD I TALK TO MY CHILD?



When a child is sick, parents and carers tend to become overprotective or sometimes over-permissive, letting children do things they usually are not allowed to do. Children crave structure and may become scared or confused if you start changing or breaking your own rules. As much as possible, try to maintain the same family routine you had before your child became ill.

Parents and carers sometimes struggle to speak to a child about their illnesses. Share age-appropriate information, bearing in mind that older children may want to know more about their disease and treatment. Don't give too much information; children commonly process information in small amounts, which allows them to cope at their own pace. Don't try to hide the facts. If a child overhears a doctor or doesn't understand what's happening, they may begin to imagine the worst.

Talk to your child about what they are feeling. Parents may be surprised by which aspects of an illness are most difficult for a child. For instance, some children may find not being able to do things they used to do more stressful than uncertainty about their illness.

Children who are sick can also feel isolated at school. Help them practice a short script to explain their condition to friends or classmates who ask questions.

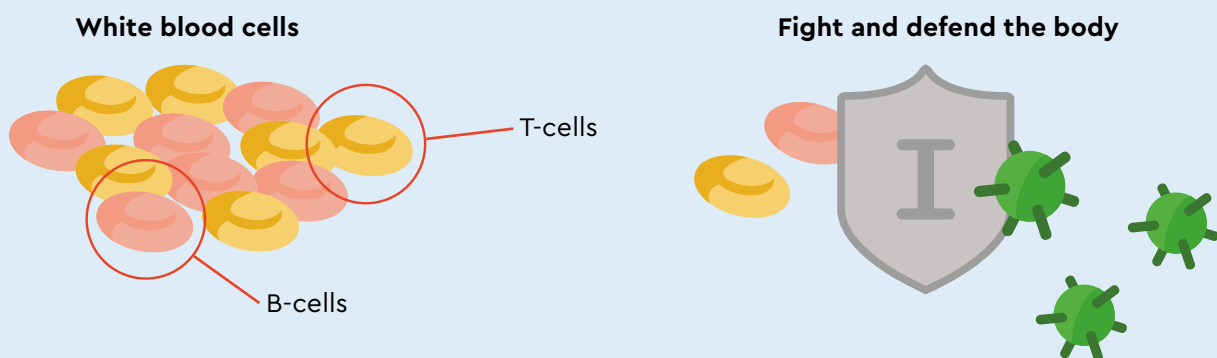
Reassure your child that it is not their fault and encourage them to ask questions or discuss their concerns. Help your child recognize that the medical care team is working to help them get well and explain the reasons for the tests and treatments.



## Explaining PTLD to your child

### What are white blood cells, and how are they made?

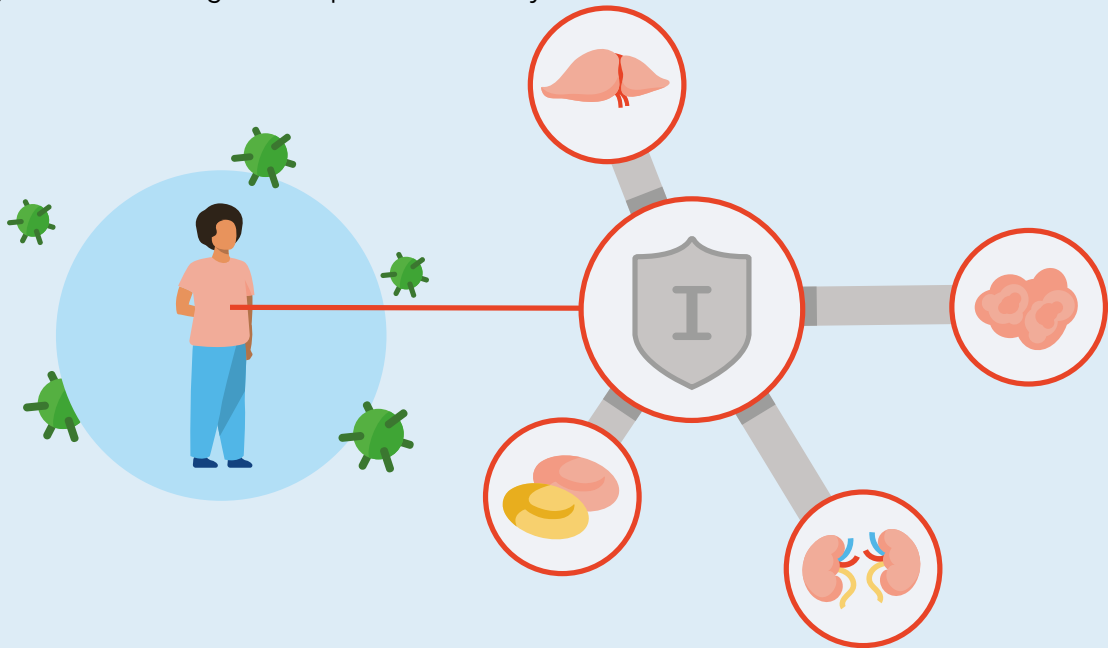
White blood cells are warrior cells in our body and part of our immune system. B-cells and T-cells are two types of white cells. They help fight and defend the body against invaders (e.g, tumor cells, infected cells).





### What does the immune system do?

To be immune means to be protected. So it makes sense that the body system that helps fight off sickness is called the immune system. The immune system is made up of a network of cells, tissues, and organs that work together to protect the body.



### The job of white blood cells

B-cells and T-cells are white blood cells in the immune system. Both are needed to help the body defend against infection and disease, but they have different jobs.

**B-cells produce antibodies against invaders**

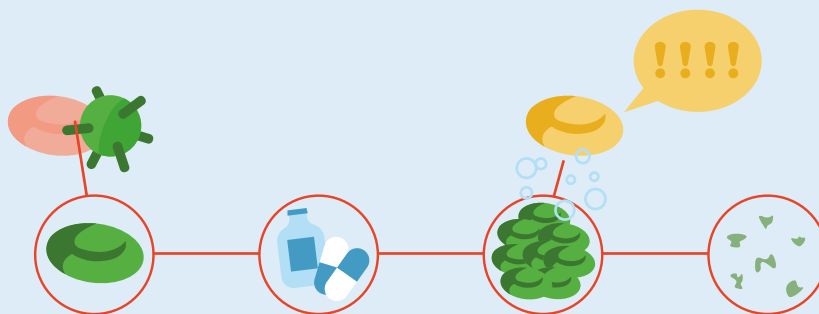


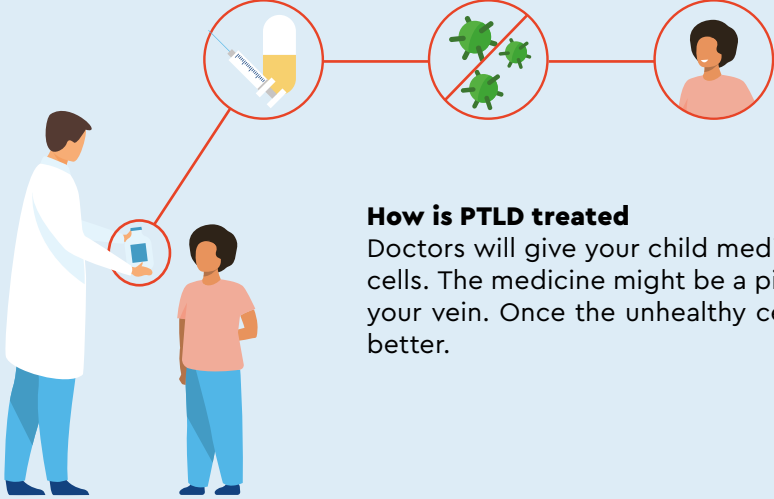
**T-cells attack invaders directly**



### What happens when you have PTLD?\*

B-cells can multiply and spread without control because of the immunosuppressant drugs used after transplantation.





**How is PTLD treated**  
Doctors will give your child medicine to get rid of your unhealthy cells. The medicine might be a pill, injection, or intravenously into your vein. Once the unhealthy cells are gone, your child will feel better.

## WHAT OTHER SUPPORT IS AVAILABLE TO ME?

In addition to support from family and friends, connecting with other PTLD patients and parents or carers of children with PTLD through patient support groups can be extremely valuable. Sharing experiences and feelings with people who know what it's like to look after a child with PTLD can be comforting and empowering.

You could contact several organizations and patient support groups following a PTLD diagnosis for more support and advice.

**The following is a list of European and International Patient Support Groups:**

- **Lymphoma Coalition**  
[www.lymphomacoalition.org](http://www.lymphomacoalition.org)





# GLOSSARY OF MEDICAL TERMS

**Allogeneic:** An allogeneic stem cell transplant uses healthy blood stem cells from a donor to replace the unhealthy bone marrow of a recipient. An allogeneic stem cell transplant is also called an allogeneic bone marrow transplant.

**Antibody:** Antibodies are disease-fighting proteins in the body that play a crucial role in the immune system.

**B-cells:** A type of white blood cell that makes antibodies. B-cells are part of the immune system and develop from stem cells in the bone marrow.

**B-cell lymphoma:** A type of cancer originating from B-cells (immune system cells). B-cell lymphomas may be either indolent (slow-growing) or aggressive (fast-growing). Most B-cell lymphomas are non-Hodgkin lymphomas. There are many different types of non-Hodgkin B-cell lymphomas.

**Bone marrow:** A soft, spongy area in the center of some of the body's larger bones. It produces all the different cells that make up the blood, such as red blood cells, white blood cells (of many different types), and platelets. All of the cells of the immune system are also made in the bone marrow.

**Chemo-immunotherapy:** Chemotherapy combined with immunotherapy. Chemotherapy uses different drugs to kill or slow the growth of cancer cells; immunotherapy uses treatments to stimulate or restore the ability of the immune system to fight cancer.

**Epstein-Barr virus (EBV):** EBV is a common virus that remains dormant in the person's body once infected. It is a lifelong infection. EBV causes infectious mononucleosis and has been associated with certain cancers, including Burkitt lymphoma, immunoblastic lymphoma, nasopharyngeal cancer, and stomach (gastric) cancer. Also called the Epstein-Barr virus.

**EBV<sup>+</sup> PTLD:** Post-transplant lymphoproliferative disease (PTLD) is a life-threatening complication of solid organ transplantation (SOT) and hematopoietic cell transplantation (HCT). In most cases, PTLD is associated with active replication of Epstein-Barr virus (EBV) after either primary infection or reactivation during treatment with immunosuppressive drugs.

**Haemato-oncologist:** A hematologist-oncologist is a physician who specializes in diagnosing, treating, and/or preventing blood diseases and cancers.

**HCT:** Hematopoietic cell transplantation (also called bone marrow transplantation or stem cell transplantation) is a type of treatment for cancer (and a few other conditions as well).

**Lymph nodes:** Lymph nodes are small, oval-shaped organs that contain immune cells to attack and kill foreign invaders, such as viruses.

**Lymphoma:** Lymphoma is a broad term for cancer that begins in cells of the lymph system. The two main types are Hodgkin lymphoma and non-Hodgkin lymphoma (NHL).

**PTLD:** A condition in which lymphocytes (usually B cells) grow out of control after transplantation in patients with weakened immune systems. This usually happens if the patient has also been infected with the Epstein-Barr virus. Also called a post-transplant lymphoproliferative disorder.

**SOT:** Solid organ transplantation is a treatment option for end-stage organ failure of the kidneys, liver, pancreas, heart, and lungs. Transplantation involves the removal of an organ from a donor to place it in the recipient's body. The new organ replaces a damaged one.

**T-cells:** A type of white blood cell. T-cells are part of the immune system and develop from stem cells in the bone marrow. They help protect the body from invaders (viruses, cancer cells).



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LABORATOIRES

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