Plasma cell leukaemia

Related conditions Infosheet

This Infosheet provides information about plasma cell leukaemia (PCL), how it is diagnosed, and how it is treated.

What is PCL?

Plasma cell leukaemia (PCL) is a rare type of cancer. In plasma cell leukaemia, a type of **white blood cell**, called a **plasma cell**, becomes abnormal and starts to rapidly multiply out of control. These abnormal plasma cells are normally made and found in the spongy centre of bones called the **bone marrow**. In PCL, these abnormal plasma cells then get into the blood.

PCL is similar to another cancer called **myeloma.** It is generally considered to be an advanced form of myeloma. However, in myeloma most of the abnormal plasma cells stay within the bone marrow. PCL is considered more aggressive and fast changing than myeloma. It normally requires intensive treatment and monitoring to start quickly after diagnosis.

There are two types of PCL:

- **Primary PCL –** diagnosed in people with no history of myeloma
- **Secondary PCL –** diagnosed after a previous diagnosis of myeloma. Occurs when myeloma changes to PCL, often after many treatments and relapses

For more information about myeloma see **Myeloma – An introduction** from Myeloma UK

How common is PCL?

Both primary and secondary PCL are rare. Each year, fewer than 1 in 1 million people will develop primary PCL.

Secondary PCL develops in between 1 in 100 and 4 in 100 people of all people with myeloma. The number of people developing secondary PCL has increased over recent years. This is thought to be because more people are living longer with their myeloma.

What causes PCL?

The exact causes of PCL are not known. It is thought to involve a series of complex changes to a person's genetics. However, what triggers these changes is unknown.

There are some factors that may increase the risk of developing PCL including:

- Age: Primary PCL is most commonly diagnosed aged 55 to 60 years. Secondary PCL is commonly diagnosed age 65 to 70 years
- Gender: PCL is slightly more common in men than women
- Exposure: Some industrial and environmental factors can increase risk of PCL

What are the symptoms and complications of PCL?

The initial symptoms and complications of primary PCL are similar to myeloma. However, they are normally more severe. Symptoms can include:

- Anaemia
- Bone pain or breaks
- Fatigue
- Recurrent infections
- Bleeding or bruising easily
- Hypercalcaemia (high blood calcium levels)
- Kidney damage
- Enlarged liver, spleen, or lymph nodes

How is PCL diagnosed?

PCL is diagnosed by the number of abnormal plasma cells in the blood. Primary PCL is diagnosed when there are symptoms and when at least 5 in 100 white blood cells are abnormal plasma cells.

Secondary PCL is diagnosed when there is a history of myeloma and when at least 20 in 100 white blood cells are abnormal plasma cells.

Some hospitals may diagnose secondary PCL when there is a history of myeloma and when at least 5 in 100 white blood cells are abnormal plasma cells. However, this is not routinely done throughout the UK.

A range of other tests and investigations will happen to diagnose PCL including:

- Blood and urine tests
- Bone marrow aspiration and biopsy
- Scans such as MRI or PET-CT

Many of these tests and investigations are also used to diagnose myeloma.



For more information, see the **Tests and investigations in myeloma Infoguide** from Myeloma UK

How is PCL treated and managed?

Treatment will depend on the type of PCL you have.

Primary PCL

Treatments for primary PCL are similar to those for myeloma. Primary PCL is generally more aggressive than myeloma, and patients are likely to have more complications when they are diagnosed. For this reason, treatment normally starts as quickly as possible.

Like myeloma, primary PCL is a relapsing and remitting cancer. This means that following treatment, there will be periods of **remission,** where the signs and symptoms of PCL are gone or at a very low level. This is followed by **relapse** where the PCL becomes active again.

Your treatment will depend on the stage of your primary PCL (newly diagnosed or relapsed), and on your age and overall fitness. Different combinations of the following types of treatment may be used:

- Proteasome inhibitor such as bortezomib (Velcade®), carfilzomib (Kyprolis®), and ixazomib

- Immunomodulatory drug (IMiD) such as lenalidomide (Revlimid®), thalidomide ,and pomalidomide (Imnovid®)
- Steroid such as dexamethasone and prednisolone
- Monoclonal antibody such as daratumumab (Darzalex®)
- Chemotherapy such as cyclophosphamide and doxorubicin



For more information see the **Treatment Guides** from Myeloma UK

Some younger and fitter people may be offered **high-dose therapy and autologous stem cell transplant (HDT-SCT).** This is an intensive treatment that involves using your own blood stem cells. Stem cells are the body's building blocks, with the ability to transform into different types of blood cell.

A HDT-SCT involves:

- Induction treatment: Treatment given over several months to try and reduce the number of abnormal plasma cells in your body
- Stem cell collection and storage: Your own stem cells are collected from your blood and stored
- High dose therapy: High doses of a chemotherapy called melphalan are given, to kill as many as possible of the abnormal plasma cells in the bone marrow
- Stem cell transplant: Your stored stem cells are put back into the blood where the bone marrow recovers and the stem cells start to make new healthy blood cells
- Maintainence treatment: Treatment given over a longer period of time to try and lengthen remission time after transplant

Some people may have a second HDT-SCT soon after their first one, or when their PCL relapses.



For more information see the **High-dose therapy and autologous stem cell transplantation Infoguide** from Myeloma UK

In a few younger and fitter people, HDT-SCT using stem cells from a donor, such as a close relative, may be considered. This is called **allogeneic HDT-SCT**. Allogeneic HDT-SCT may benefit some people but also carries extra risks.



For more information see the **Allogeneic stem cell transplant in myeloma Infosheet** from Myeloma UK

Secondary PCL

Most people with secondary PCL will already have had several myeloma treatments and some may be resistant (**refractory**) to them. Treatment options are determined on an individual basis. You may be given a combination of treatments mentioned above on page 4. You may also be offered a more intensive chemotherapy combination including cyclophosphamide, doxorubicin, vincristine, etoposide, and cisplatin, together with the steroids dexamethasone or prednisolone.

Clinical trials

Some people may be offered treatments that are still being developed. This will be on an individual basis, or as part of a clinical trial. This option may strongly be considered for people whose PCL has become refractory to other treatments. Your healthcare team will discuss these options with you.



For more information see the **Horizons Infosheets** and the **Clinical trials and how treatments are developed Infoguide** from Myeloma UK

Supportive treatment

Supportive treatments are important to help prevent or reduce the symptoms and complications of PCL. These may include painkillers, blood transfusions, antibiotics, and antiviral drugs. You will be advised to drink plenty of fluids to help with your kidney function.

Current research in PCL

Because PCL is such a rare cancer, it has been difficult to run clinical trials of different treatments. There is a need for more research.

Some clinical trials have included people with PCL alongside people with relapsed and refractory or high-risk myeloma. There have also been some studies of all people with PCL from a patient registry, to assess what treatments they were given

and what the outcomes were. These studies can provide useful information in conditions such as PCL where clinical trials are hard to do.

Newer methods of genetic sequencing are leading to a better understanding of the genetic changes in plasma cells in people with PCL.

Genetic studies of PCL may eventually increase our understanding of how it develops and progresses, and who is at higher risk of getting PCL.

Living well with PCL

Hearing that you have a rare cancer can bring worry and anxiety for yourself and those you are close to. There is no right or wrong way to feel about your diagnosis.

The consequences of living with PCL can be wide-ranging and may affect areas of your life that you were not expecting to change. Understanding and being aware of any changes and how they may make you feel can help you to work through them.

Some of the changes may be harder to adjust to than others, and if you are struggling, there is support and advice available to you. Speak to your healthcare team or GP as they can explore ways to help you manage.

Sources of support

It is normal to have questions about PCL or want to talk with someone who understands your situation. You can access support from:

- Your healthcare team – if you are concerned about your treatment or any symptoms, speak to your healthcare team
- The Myeloma UK Infoline – call **0800 980 3332** or email **AskTheNurse@myeloma.org.uk**. Our team of specialists can provide emotional support and practical information
- Family and friends – who you tell about your diagnosis, and when, is a personal decision. However, the friends or family that you do tell can be an important source of support
- A trained Myeloma UK Peer Buddy, matched with you – chatting with someone else who has PCL can help
- The Myeloma UK Discussion Forum – go to **forum.myeloma.org.uk** to talk with others affected by PCL

- Counselling or other talking therapies – these may help if you are finding things challenging. Your healthcare team can give you information on available services

Myeloma UK is here for everything a diagnosis brings. Get in touch to find out more about how we can support you by calling the Infoline on **0800 980 3332** or visiting our website **myeloma.org.uk**

Key points

- Plasma cell leukaemia (PCL) is a rare cancer
- A type of white blood cell, called plasma cells, become abnormal and some leave the bone marrow and get into the blood
- PCL is different to myeloma where most of the plasma cells remain in the bone marrow
- PCL may develop as a late stage of myeloma (secondary PCL), or occur in patients without previous myeloma (primary PCL)
- The causes of PCL are not known
- Symptoms and complications can include anaemia, bleeding easily, bone pain, fatigue, recurrent infections, and kidney damage
- PCL is generally more aggressive than myeloma. The range of treatment options is similar to that in myeloma
- Help and support is available for you. Speak to your healthcare team or Myeloma UK for more information

Medical terms explained

Bone marrow: The soft, spongy tissue in the centre of bones that produces blood cells.

High-dose therapy and allogeneic stem cell transplant (Allogeneic HDT-SCT): Treatment with high doses of chemotherapy given into a vein, before patients receive a donors healthy stem cells as part of the stem cell transplant procedure.

High-dose therapy and autologous stem cell transplant (HDT-SCT): Treatment with high doses of chemotherapy given into a vein, before patients receive their own healthy stem cells as part of the stem cell transplant procedure.

Myeloma: A cancer of the bone marrow caused by abnormal plasma cells which results in bone damage, low blood cell counts, increased infections, and kidney damage.

Plasma cells: A type of white blood cell that produces antibodies to fight infection.

Plasma cell leukaemia (PCL): A rare and aggressive cancer of the plasma cells, with plasma cells found in the blood. It can start itself or can evolve from advanced myeloma.

Refractory: Disease that has failed to respond to a treatment or has stopped responding to the treatment.

Relapse: The point where disease returns or becomes more active after a period of remission or plateau.

Remission: The period following treatment when plasma cells are no longer detectable, or at a very low level, and there are no clinical symptoms of plasma cell leukaemia.

White blood cell: A type of blood cell involved in the body's immune system, which helps to fight infection and disease.

About this Infosheet

The information in this Infosheet is not meant to replace the advice of your healthcare team. They are the people to ask if you have questions about your individual situation.

For more about how we produce our information, visit **myeloma.org.uk/about-our-information**

We value your feedback about our patient information. For a short online survey go to **myeloma.org.uk/pifeedback** or email comments to **patientinfo@myeloma.org.uk**

Other information available from Myeloma UK

Myeloma UK has a range of information booklets available covering all aspects of myeloma and related conditions. Download or order them from **myeloma.org.uk/publications**

To talk to one of our Myeloma Information Specialists about any aspect of myeloma, call our Myeloma Infoline on **0800 980 3332** or **1800 937 773** from Ireland.

The Infoline is open from Monday to Friday, 9am to 5pm and is free to phone from anywhere in the UK and Ireland.

Information and support about myeloma is also available at **myeloma.org.uk**

Related conditions Infosheet:
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MyelomaUK

We're here for everything a diagnosis of plasma cell leukaemia brings

Get in touch to find out more about how we can support you

Call the Myeloma Infoline on
0800 980 3332

Email Ask the Nurse at
AskTheNurse@myeloma.org.uk

Visit our website at
myeloma.org.uk

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