

Plasma Cell Leukaemia

This Infosheet explains what plasma cell leukaemia is, how it is diagnosed, and how it can be treated and managed.

What is PCL?

Plasma cell leukaemia (PCL), also known as plasma cell myeloma, is a rare type of cancer. Similar to myeloma, PCL affects the plasma cells that are normally found in the bone marrow and form part of the immune system. In myeloma and PCL, the plasma cells become abnormal and multiply out of control.

PCL is considered to be a more advanced form of myeloma and usually requires more intensive monitoring and treatment.

In PCL, a significant proportion of the abnormal plasma cells leave the bone marrow and are released into the blood. This is unlike myeloma, where the majority remain in the bone marrow.

There are two types of PCL:

- **Primary PCL** – diagnosed in patients with no history of myeloma
- **Secondary PCL** – occurs after a previous myeloma diagnosis and arises when myeloma progresses to PCL

The likelihood of myeloma patients developing secondary PCL is very low (about 0.5 - 1%). Secondary PCL is thought to be caused by an accumulation of specific genetic changes that occur within myeloma cells as the myeloma progresses in certain patients. However, at present, little is understood about the genetic differences between myeloma and PCL and there is no way of predicting which myeloma patients will progress to PCL.

Who develops PCL?

PCL is rare. It is estimated that 1 per million of the general population are diagnosed with primary PCL each year, while approximately 1 in 100 myeloma patients will go on to develop secondary PCL. The average time for myeloma to develop into secondary PCL is approximately 21 months.

The average age of diagnosis of primary PCL is 55 years old, which is slightly younger than in myeloma. The average age of patients diagnosed with secondary PCL is 66 years old.

Both primary and secondary PCL are slightly more common in men than in women, and in the Afro-Caribbean and African populations than in the Caucasian population.

What causes PCL?

The exact causes of PCL are not fully understood. Like myeloma, a series of genetic changes during the development of a plasma cell is thought to lead to its uncontrolled growth. However, what triggers these changes is not known. Exposure to additional risk factors is thought to play an important role, including age and exposure to industrial and environmental factors.

What are the symptoms and complications of PCL?

PCL patients normally present with similar symptoms and complications to myeloma; however, they tend to be more severe.

Symptoms and complications include:

- Anaemia
- Bone pain
- Fatigue
- Recurrent infections
- Bleeding
- Hypercalcaemia (high blood calcium levels)
- Kidney damage

Patients may also have an enlarged liver or spleen caused by a large number of abnormal plasma cells accumulating and being deposited in these organs. This is more common in primary than in secondary PCL.

How is PCL diagnosed?

PCL is diagnosed by the number of abnormal plasma cells in the blood. A diagnosis is confirmed when there are more than 2 million abnormal plasma cells per millilitre of blood, or when abnormal plasma cells make up more than 20% of the total number of white blood cells present in the blood.

How is PCL treated and managed?

Current treatments for primary PCL are the same as those used in myeloma and include drugs

such as thalidomide, bortezomib (Velcade®) and lenalidomide (Revlimid®). High-dose therapy and stem cell transplantation is generally offered to younger and/or fitter patients.

Most secondary PCL patients will have already had several anti-myeloma treatments and some patients may have become refractory (resistant) to them. For these patients, newer drugs such as daratumumab (Darzalex®) or more intensive treatments combinations using chemotherapy drugs including cyclophosphamide, doxorubicin, vincristine, etoposide and cisplatin, together with the steroids dexamethasone or prednisolone, may be considered.

Supportive treatment is important to help prevent or reduce the symptoms and complications of PCL. These may include pain-killers, blood transfusions and antibiotics.

Future directions

It is hoped the development of novel treatments for myeloma will also benefit PCL patients. Newer drugs such as carfilzomib (Kyprolis®) and pomalidomide (Imnovid®), and drugs with entirely new mechanisms of action such as daratumumab, have been

shown to be effective in myeloma patients who no longer respond to bortezomib and lenalidomide and could therefore also provide new treatment options for PCL patients.

About this Infosheet

The information in this Infosheet is not meant to replace the advice of your medical team. They are the people to ask if you have questions about your individual situation. All Myeloma UK publications are extensively reviewed by patients and healthcare professionals prior to publication.

Other information available from Myeloma UK

Myeloma UK has a range of Essential Guides, Infoguides and Infosheets available covering many areas of myeloma, its treatment and management. To order your free copies or to talk to one of our Myeloma Information Specialists about any aspect of myeloma, call the **Myeloma Infoline: 0800 980 3332** or **1800 937 773** from Ireland

The Myeloma 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 around the clock at www.myeloma.org.uk

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