

# POEMS syndrome

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**This Infosheet explains what POEMS syndrome is, how it is diagnosed and how it is treated and managed.**

## **What is POEMS syndrome?**

POEMS syndrome is a rare type of plasma cell disorder that can affect multiple systems in the body. It is named after the five common features of the syndrome, described below:

- **Polyneuropathy** – nerve damage to various peripheral nerves which control muscles and sensation. Also known as peripheral neuropathy
- **Organomegaly** – enlargement of organs, such as the liver, spleen or lymph nodes
- **Endocrinopathy** – abnormal function of endocrine (hormone-producing) glands
- **Monoclonal plasma cell disorder** – such as myeloma, MGUS (see below) and other plasma cell disorders
- **Skin changes** – certain skin changes are characteristic in POEMS syndrome, such as hyperpigmentation (darkening of an area of the skin or nails)

These five features - known as the 'major criteria' of the disorder - are the main characteristics of POEMS syndrome. There are other possible features of POEMS syndrome ('minor criteria') not included in the acronym which are:

- Abnormal bone growth in certain parts of the skeleton. They are typically painless but are usually visible on X-rays and scans
- Swelling of the optic nerve, the main nerve in the eye (known as papilloedema)
- Fluid build-up around the lungs, in the abdomen and/or the legs
- High red blood cell levels and/or high platelet levels in the blood
- Raised levels of a cytokine (chemical messengers produced by some immune system cells) in the body known as vascular endothelial growth factor (VEGF)
- Deep vein thrombosis - the formation of a blood clot that occurs in the deep veins of the body, typically the legs
- Poor blood circulation
- Breathlessness or difficulty breathing

POEMS syndrome is also known as osteosclerotic myeloma, Takatsuki syndrome and Crow-Fukase syndrome, however these terms are less commonly used.

### What is a monoclonal plasma cell disorder?

A plasma cell disorder is the term used to describe a condition which produces abnormal plasma cells. Plasma cells are a type of white blood cell that produce antibodies (immunoglobulins). Antibodies bind to substances in the body that are recognised as foreign, such as bacteria and viruses (known as antigens), enabling other cells of the immune system to destroy and remove them.

In plasma cell disorders, the abnormal plasma cell produces an abnormal antibody known as paraprotein (also called monoclonal or M protein) which has no useful function.

The most common plasma cell disorders associated with POEMS syndrome are:

- Monoclonal gammopathy of undetermined significance (MGUS) - a non-cancerous condition in which low levels of

paraprotein are present in the blood and/or urine. Patients do not have symptoms but have an increased risk of developing myeloma

- Myeloma – a cancer arising from plasma cells that are normally found in the bone marrow. Common symptoms/ complications include bone damage, fatigue, increased infection and kidney damage

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For more information see the **MGUS Infosheet** and **Myeloma – an Introduction** from **Myeloma UK**.

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### What causes POEMS syndrome?

The exact causes of POEMS syndrome are not well understood.

The paraprotein produced by the abnormal plasma cells is on its own not enough to explain the many features of POEMS syndrome. Various cytokines are thought to play a role in causing damage to the different tissues and organs involved in the syndrome. This includes the cytokine VEGF that is found in higher levels in POEMS syndrome patients.

### Who can develop POEMS syndrome?

POEMS syndrome is very rare and the incidence is not fully known. It is thought that many cases remain undiagnosed because of its rarity and the fact that patients may be seen by different doctors who are unfamiliar with the syndrome.

The average age of diagnosis is in people in their 50s, however this can range from 30 – 80 years old. POEMS syndrome is twice as common in men as in women and there is not thought to be any difference in incidence in different ethnic populations.

### What are the symptoms of POEMS syndrome?

The most common symptoms result from the peripheral neuropathy associated with the syndrome. Peripheral neuropathy is often the most debilitating feature of POEMS syndrome.

The first signs of peripheral neuropathy include numbness and tingling in the hands and feet which progressively worsens over time. Pain, discomfort and weakness in the hands and feet are common features of POEMS syndrome and weakness is often an early symptom of the syndrome.

Other symptoms vary depending on the organ systems involved and can include:

- Weight loss
- Diarrhoea
- Enlargement of the lymph glands (also known as lymph nodes)
- Fluid build-up in the feet and ankles
- Increased sweating
- Skin changes, including:
  - Thickening of the skin
  - Red or purple spots on the surface on the skin
  - An increasing amount of hair on the arms and legs which is often coarse in texture
- Swelling of the fingertips and white nails (known as 'clubbing')
- Headaches or blurred vision
- Reduced sexual function (reduced libido, loss of erections)

Patients may have fewer symptoms at diagnosis but develop more symptoms over time as the syndrome progresses.

## How is POEMS syndrome diagnosed?

Not all of the five major criteria (polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder and skin changes) are always present in POEMS syndrome. POEMS syndrome may be diagnosed if two major criteria and one additional minor criteria are present in a patient.

To confirm a diagnosis, your doctor will need to perform a thorough physical examination where they will look for:

- Skin and hair changes
- Evidence of fluid build-up
- Enlargement of specific glands and organs such as lymph glands, liver and/or the spleen
- Signs of optic nerve swelling
- Signs of neuropathy by conducting a complete examination of the nervous system

Your doctor will confirm a diagnosis through a number of tests and investigations which include:

**1.** Blood tests, which check:

- Your levels of white blood cells, red blood cells and platelets

- Your liver and kidney function
  - If paraprotein is present
  - The level of certain cytokines such as VEGF
2. Your urine may also be tested for the presence of part of the paraprotein called light chains (also sometimes called Bence Jones Protein)
  3. X-rays and scans to look for abnormal bone growth or bone damage
  4. Biopsies and other tests:
    - A bone marrow biopsy, or a biopsy of any abnormal bone growth, may be performed to look for the presence of abnormal plasma cells
    - A nerve conduction test may be performed to assess nerve function and damage

### How is POEMS syndrome treated?

Treatment of POEMS syndrome will depend on several factors:

- The type of underlying plasma cell disorder (e.g. MGUS or myeloma)
- The main symptoms present and organs affected
- Age and general fitness of the patient

Treatment may include chemotherapy and/or radiotherapy.

### Radiotherapy

You will typically receive radiotherapy treatment if you have an abnormal bone growth in just one or two areas of the bone or there is evidence of solitary plasmacytoma (a discreet, single mass of abnormal plasma cells which can occur in bone or in soft tissue), but no evidence of abnormal plasma cells spread throughout the rest of your body. You will be assessed by a radiotherapy specialist who will decide on the exact amount and number of treatments needed.

### Chemotherapy

You will typically receive chemotherapy-based treatment if the abnormal plasma cells are more widely spread throughout the rest of your body. It is likely that you will receive a combination of different types of drugs which work together. The most common types of drugs used to treat POEMS syndrome are:

- An alkylating agent which is a type of chemotherapy drug, such as melphalan or cyclophosphamide

- A steroid, such as dexamethasone or prednisolone

These drugs may also be combined with drugs used to treat the underlying plasma cell disorder such as thalidomide, bortezomib (Velcade®) and lenalidomide (Revlimid®). These should be given at low doses, as both thalidomide and bortezomib can damage the peripheral nerves, possibly increasing the polyneuropathy feature of POEMS syndrome.

### **Other forms of treatment**

High-dose therapy and stem cell transplantation may be an option for some patients, especially those with many symptoms or with a rapidly deteriorating syndrome. Though data on the effectiveness of treatment for POEMS syndrome is limited, a recent retrospective study of 127 patients found high-dose therapy and stem cell transplantation to be a suitable treatment option for POEMS syndrome patients, providing good long-term outcomes.

A treatment known as plasma exchange, frequently used for other nerve disorders, is not recommended as it has been shown to be ineffective for POEMS syndrome.

Treatment is effective in the majority of patients, but improvement is normally gradual. Once there is evidence of an improvement in symptoms, quality of life and day-to-day functioning, it is recommended that patients are referred for a period of rehabilitation, preferably at a specialist rehabilitation centre, to allow nerve and muscle function to improve.

Nerve pain is common in POEMS syndrome and is often treated with drugs such as amitriptyline, gabapentin or pregabalin. Pain caused by nerve damage can be difficult to treat or manage and may require input from a pain specialist or palliative care.

### **How is POEMS syndrome monitored?**

POEMS syndrome patients should be monitored regularly and the type of treatment they receive will determine the frequency of their hospital visits.

After initial treatment is complete, patients are typically seen every three months for a physical assessment and regular blood tests. Patients are likely to receive other ongoing assessments such as nerve conduction tests and scans to assess any old or new abnormal bone growth.

## Future directions

Ongoing research aims to better understand what causes POEMS syndrome and why only some patients with certain plasma cell disorders develop the syndrome.

Research is investigating whether new drugs treating the underlying plasma cell disorder can slow down the progression of POEMS syndrome. For example some of the newer drugs used to treat myeloma such as ixazomib (Ninlaro®) are being studied for use in POEMS syndrome.

Chemotherapies used for other cancers are also being investigated for use in POEMS syndrome, such as bevacizumab (Avastin®) which is a drug that is used in several different cancers.

It targets the cytokine VEGF, which is present in higher than normal amounts in POEMS syndrome patients. It has been trialled in a small number of patients, with mixed results.

## 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. For a list of references used to develop our resources, visit [www.myeloma.org.uk/references](http://www.myeloma.org.uk/references).

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## 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](http://www.myeloma.org.uk)

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