

Your guide to the

# Waldenstrom's macroglobulinaemia guidelines

## What are the guidelines?

The British Society for Haematology publishes [guidelines](#) for the diagnosis and management of Waldenstrom's macroglobulinaemia (WM). Guidelines like these are developed to provide healthcare professionals with clear guidance on how to care for their patients.

For people with WM, this means you can expect improved care, no matter where you are being treated in the UK. Here, we've summarised the latest guidelines and what it means for your care. We hope by doing so that you will feel more confident talking with your healthcare team and in taking an active role in your care.

## Diagnosis



- If you are suspected of having WM, your doctor should first take [blood tests](#)
- A diagnosis of WM can only be confirmed by a bone marrow biopsy - both [bone marrow aspirate](#) and [trephine](#)
- You should be assessed to see if you have a mutation in the [MYD88 gene](#). Your doctor should consider you for assessment for other gene mutations

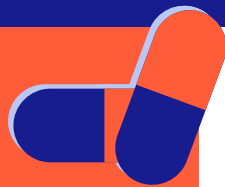
## Before treatment

- After diagnosis, it may be safe for your doctor to monitor you at 3-6 monthly intervals - known as '[Active Monitoring](#)'
- Factors that might indicate you need treatment include: symptoms that significantly impact your life, enlarged lymph nodes, developing a condition related to high IgM levels, or a low blood count

## First line treatment

- The guidelines list acceptable treatments which current evidence have proven effective
- The two most commonly used first line therapies for WM are [chemotherapy combinations](#) DRC (dexamethasone, rituximab and cyclophosphamide) and BR (rituximab-bendamustine)
- Using [rituximab](#) alone isn't recommended for the majority of people, and is saved only for frail or elderly people
- There is a lack of evidence to support the use of maintenance rituximab
- Regimens that include a type of drug called a proteasome inhibitor - [bortezomib](#) - and [BTK inhibitors](#) like ibrutinib and zanubrutinib are also acceptable as first line treatments, but aren't funded by the NHS

## Second line treatments



- Your treatment options if your WM relapses depend on what your previous treatment was and how long it's been since you had treatment
- All rituximab regimens, as well as BTK inhibitors like ibrutinib, are options for second line treatment
- Bortezomib regimens are also acceptable but not available of the NHS
- [Stem cell transplants](#) can be considered as a second line treatment if you were responsive to chemotherapy. However, this form of treatment isn't recommended to people who had less than a partial response

## Other treatment considerations

- There isn't a specific sequence of therapies recommended for people with WM
- Treatment is unique to you. Your doctor should take into account your specific symptoms, your fitness and other medical problems
- Your doctor should use set criteria to evaluate your response to treatment. However, they should also take into account other factors, for example how improved your quality of life is
- Your doctor should also consider you for [clinical trials](#)

## When WM transforms

- Rarely, WM can transform - or change - into a faster-growing form of lymphoma, often diffuse large B-cell lymphoma (DLBCL)
- If your doctor suspects your WM has transformed, you should have a tissue biopsy
- If your WM has transformed, your treatment will be similar for people who develop DLBCL without WM



## Complications & related conditions

- Healthcare professionals are encouraged to work with specialist colleagues to help treat any complications or conditions you might have e.g. neuropathy
- Doctors should seek another opinion from colleagues with expertise in WM
- Your doctor should investigate any symptoms that might be related to WM, whatever your blood test results, as it might mean you need to start treatment

## Supportive care

- You should be offered the seasonal flu vaccine, COVID-19 vaccines and pneumococcal conjugate vaccine
- Live vaccines (like polio, herpes zoster and yellow fever) are not recommended. However if you're eligible for the shingles vaccine, you can now receive a non-live vaccine called Shingrix
- If you have hypogammaglobulinaemia (low levels of IgG) and develop recurrent bacterial infections, you should be considered for preventative treatment
- If you have WM, you are classed as clinically extremely vulnerable in relation to COVID-19, whether you are on treatment or not

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