

The CLL Haematology Service: Information for GPs

One of your patients has been diagnosed with either MBL, SLL or CLL. This leaflet tells you about referrals to the Haematology Service, as well as other issues you are likely to encounter, including managing chest infections and vaccinations.

As CLL and SLL are very similar conditions, and treated in the same way, all references to CLL below also apply to SLL.

These are general guidelines and there may be regional variations.

Please add this leaflet to your patient's home page for continued ease of access

Definition of MBL, SLL and CLL

CLL arises from a pre-malignant condition called **monoclonal B-cell lymphocytosis** (MBL). High-count MBL is defined as the presence of a clonal B-cell population of greater than $0.5 \times 10^9/l$ but less than $5 \times 10^9/l$ when tested by flow cytometry.

Individuals with MBL have an annual progression risk to Stage A CLL of approximately 4%.

It is important to perform a clinical examination for lymph node and splenomegaly in all patients. If present, in patients with a circulating CLL-phenotype B cell clone $<5 \times 10^9/l$, a diagnosis of **small lymphocytic lymphoma** (SLL) is made, which is managed in the same way as CLL. Patients with a circulating CLL-phenotype B cell clone $>5 \times 10^9/l$, whether there is palpable lymphadenopathy or not, meets diagnostic criteria for **chronic lymphocytic leukaemia**.

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CLL is staged as follows:

Stage A	Less than three groups of enlarged lymph nodes* No anaemia/thrombocytopenia
Stage B	More than three groups of enlarged lymph nodes* No anaemia/thrombocytopenia
Stage C	Anaemia and/or thrombocytopenia

Fig 1 Binet Staging System for CLL patients.

**As assessed clinically*

How to refer to your NHS CLL Haematology service

Individuals with isolated lymphocytosis (MBL or Stage A CLL) should be referred as routine to the haematology clinic.

Patients with stage B or C CLL, particularly those with ALERT symptoms (see below) should be referred as URGENT.

In many regions, stable stage A patients may be followed up via telephone consultation, with bloods beforehand, often lead by Clinical Nurse Specialists.

In some regions, stage A patients may be discharged for follow-up in primary care with prior agreement with the ICB.

Stage B and C patients will generally be reviewed in the CLL clinic.

Progression of CLL

For patients with known CLL, please update secondary care team if any of the ALERT features below occur:

Progressive falling blood counts (Alerts to be sent by the Haematology Laboratory)	Haemoglobin < 100 g/L Platelets < 100 x 10 ⁹ /L
Rising Lymphocyte Count	Lymphocyte count > 50 x 10 ⁹ /L and <u>lymphocyte count doubling ≤ six months</u>
New swelling/lump	
B-symptoms	Fever of unknown cause. Drenching night sweats. Persistent new fatigue. Weight loss ≥ 10% in six months

Fig 2 ALERT symptoms of progressive CLL of any stage that require referral to specialist CLL Team

Management of chest infections

Patients with CLL are prone to chest infections and development of secondary bronchiectasis. Most patients with CLL will develop secondary immune defects including secondary hypogammaglobulinaemia.

Recurrent chest infections are managed in conjunction with Immunology and Respiratory services.

1st line	Co-amoxiclav 625 mg for 14 days
2nd line	Clarithromycin 500 mg for 14 days
Recurrent/unresolved chest infections	Please discuss with haematology team (see contact details below)

This guidance may be modified to account for the patient's personal history, e.g. allergies, previous infections, and local antibiotic guidelines.

Skin and other secondary cancers

CLL patients are at greater risk of non-melanoma skin cancers, and should be encouraged to avoid direct sun exposure, and report any changes in their skin. There is also a risk of second malignancy in general. Regular screening is advised.

Vaccinations

A hallmark of CLL is progressive immunodeficiency, characterised by impaired responses to vaccination, including influenza, pneumococcal, and varicella zoster virus (VZV), hence the importance of instigating the schedule of vaccinations below as soon after diagnosis as possible:

Vaccination programme for CLL patients

Vaccine	When to give	Notes
PNEUMOCOCCAL VACCINES PREVENAR® PPV13 (The pneumococcal conjugate vaccine).	At diagnosis OR as a catch-up dose.	The catch-up dose is for patients who have previously only been vaccinated with Pneumovax II®.
PNEUMOVAX II® PPV23 (The pneumococcal polysaccharide vaccine).	2 months after Prevenar® Refer to Green Book Chapter 25, page 8	Irrespective of any previous vaccinations.
SEASONAL FLU VACCINE	Annually	Other members of household, including healthy children, should ideally also be vaccinated annually. Where a child receives the live nasal vaccine, they should avoid close contact with the patient for the following week*.
SHINGLES VACCINE The recombinant varicella vaccine SHINGRIX does not contain any live virus and can safely be given to people with CLL.	Two doses, 2–6 months apart	All patients with CLL aged over 50 are eligible in England and recommended even if the patient has already had shingles. N.B. The live shingles vaccine (Zostavax) must NOT be used
COVID VACCINE Current approved COVID-19 vaccines	According to current UK/ Devolved Nations Government Schedule	We recommend all CLL patients to have the vaccine. CLL patients are in the “clinically extremely vulnerable group” regardless of their stage of CLL. For more information, see overleaf.

**While there is a theoretical risk of transmission of the virus from a child receiving the nasal influenza vaccine to an immunocompromised person, there have been no reported cases of illness or infections from the vaccine virus. However, as a precaution, patients should avoid contact with children for one week after the live vaccine.*

COVID-19 Vaccination

In addition to all patients keeping up to date with their vaccinations we recommend household contacts of CLL patients also be vaccinated against COVID-19 to minimise the risk of transmission.

All CLL patients, regardless of stage or treatment status, should be considered for COVID-19 anti-viral or monoclonal antibody therapy if they have a positive lateral flow test. More information can be found here: <https://bloodcancer.org.uk/support-for-you/coronavirus-covid-19/>

CLL patients should NOT receive live or attenuated (weakened) viruses.

These include:

- a. Yellow fever
- b. Oral Polio Measles, mumps and rubella
- c. The live shingles vaccine (Zostavax)

Foreign travel

Please be aware of vaccination requirements if the patient is seeking to travel abroad and avoid live vaccination.

Vaccinations for patients receiving/recently received treatment.

Vaccination timings for patients who are on or have recently completed treatment will be advised by the treating Haematologist.

General health advice

The treating Haematologist may recommend that CLL patients who have had shingles should receive lifelong Aciclovir as secondary prophylaxis once initial treatment has been successfully completed.

Patients should be encouraged to stop smoking.

CLL patients are at increased risk of UV-related skin damage and skin cancers. They should be encouraged to wear sunscreen daily with at least an SPF30 and a 5-star UVA rating.

Emotional Support — Support Act

Patients and carers/supporters can struggle with living with CLL, especially during the uncertain period of Active Monitoring/‘Watch and Wait’, or as the need for treatment approaches. A digital tool using behavioural Acceptance and Commitment Therapy (ACT) has been designed to support patients’ emotional wellbeing:

<https://cllsupport-act.org.uk/>

Useful links

CLL Support:

<http://www.cllsupport.org.uk>

Blood Cancer UK:

<https://bloodcancer.org.uk/>

Leukaemia Care:

<http://www.leukaemiacare.org.uk/chronic-lymphocytic-leukaemia>

Macmillan:

<https://www.macmillan.org.uk/cancer-information-and-support/leukaemia/chronic-lymphocytic-leukaemia-ctl>

Cancer Research UK:

<http://www.cancerresearchuk.org/about-cancer/chronic-lymphocytic-leukaemia-ctl>

The Haematology Service (for you to complete if applicable):

Contact information

CLL clinical nurse specialist: _____

Contact for re-referral or any queries: _____

Haematology registrar: _____

Information for GPs on managing and supporting patients living well with high-count Monoclonal B-cell Lymphocytosis or CLL. Information based on current BSH/UKCLL Forum guidelines. Original Authors: Dr Toby Eyre, Dr Niamh Appleby and Lianne Palmer. Additional input from Dr Sue Howarth, Dr Alison McCaig and Dr Justine Foster. Adapted for national use by CLL Support. This is a controlled document and should not be changed.

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