PRIMARY CARE MANAGEMENT GUIDELINE

# Chronic Lymphocytic Leukaemia

Date: 1 March 2005
Revision date: 1 March 2007

**Chronic Lymphocytic Leukaemia (CLL) is the most common type of leukaemia with a peak incidence between 60 and 80 years of age. Early disease has a median survival of > 12 years and often no treatment is required.**

| CLINICAL PROBLEM | ACTIONS | IMPLEMENTATION |
|---|---|---|
| **Incidental finding of peripheral lymphocytosis – diagnosis of CLL possible** | If single absolute count > or = 10 or < 10 but persists for 3 months:<br>• Full clinical examination<br>• Cell marker studies (immunophenotyping) *(note 1)* | Discuss with Haematologist if:<br>• Cell marker studies require further interpretation<br>**or**<br>• Persistent unexplained lymphocytosis |
| **Diagnosis CLL confirmed** *(note 2)* | • Clinical assessment for fever, persistent night sweats, weight loss, frequent infections, lymphadenopathy, hepatomegaly, splenomegaly<br>• Direct Coombs test<br>• Immunoglobulins<br>• Determine disease stage *(note 3)* | Refer to Haematology if any of the following:<br>• Stage A with symptoms<br>• All Stage B and C<br>• All <50 yrs irrespective of stage (may be candidates for innovative treatment strategies e.g. bone marrow transplant)<br>Discuss with Haematologist if +ve Coombs test<br>Asymptomatic Stage A patients may be managed in the community *(note 4)* |
| **Asymptomatic Stage A CLL** | • Discuss diagnosis and prognosis with patient *(note 3 and 5)*<br>• 6-monthly review including CBC, symptoms of fever, persistent night sweats, weight loss, history of recurrent infections, examination for lymphadenopathy, hepatosplenomegaly<br>• Annual Flu vaccination<br>• Consider pneumovax if coexistent chronic respiratory disorder<br>• Serum immunoglobulins if recurrent infections<br>• Regular surveillance for skin cancer which is significantly more common in association with CLL | Refer to Haematology if any of the following:<br>• Patient develops symptoms or three or more areas of organ enlargement *(note 3 and 6)*<br>• Haemoglobin falls below 100 g/L for no obvious unrelated reason or an absolute reduction in haemoglobin of >30 g/L<br>• Platelet count <100 x $10^9$/L<br>• WCC count doubles (and the total count is >30 x $10^9$/L) in less than a year<br>• Haemolytic anaemia develops<br>• Age less than 50 (may be candidates for innovative treatment strategies e.g. bone marrow transplant)<br>• Hypogammaglobulinaemia associated with recurrent infections |

Waikato District Health Board

# NOTES

**1** Both blood film examination and cell marker studies are required in order to differentiate reactive lymphocytosis and lymphomas in leukaemic phase from CLL. Bone marrow examination is not usually necessary.

**2** In the early stages, the condition is asymptomatic, the only feature being a peripheral lymphocytosis. With progression, increasing lymphocytosis, lymphadenopathy, splenomegaly and bone marrow involvement with cytopenias usually occur. Other organs are affected in some patients with late stage disease. Systemic symptoms (fever, weight loss and night sweats) are rare. Other complications of CLL include autoimmune haemolytic anaemia, immune thrombocytopenia and hypogammaglobulinaemia with recurrent infections and herpes zoster. CLL can also transform to high-grade lymphoma (Richter's transformation).

**3** Prognosis depends largely on disease stage (extent of disease). The most widely used staging system is that described by Binet:

| ***Stage*** | | ***Prognosis (Median Survival Years)*** |
|---|---|---|
| A | 0 – 2 areas of organ enlargement | > 12+ years |
| B | 3 – 5 areas of organ enlargement | 7 years |
| C | Hb < 100 g/L, or platelets < 100 x $10^9$/L (unless due to immune mechanisms) | 2 years |

The following are considered as one area of organ enlargement <u>each</u>: neck, axillae, groins, liver, spleen e.g. bilateral cervical nodes = one site

**4** Studies have demonstrated no advantage in treating asymptomatic Stage A CLL. Patients with Stages B and C disease live longer if they receive treatment and gain a response to it. Prompt treatment of infection is important for all CLL patients regardless of stage.

**5** Patient information leaflets are available via:

Haematology Outpatient Clinic, Waikato Hospital
<u>and</u>

Leukaemia & Blood Foundation of NZ
17 Kipling Ave
Auckland
PO Box 99 182
Newmarket
Auckland
09 638 3556
0800 15 10 15
Email: lbf@leukaemia.org.nz
URL: www.leukaemia.org.nz

**6** Lymphadenopathy in CLL is usually bilateral, soft, mobile and painless. Unilateral lymphadenopathy is unusual in CLL and may require further investigation.