# A Case Report of Chronic Lymphocytic Leukemia in a Young Black South African Man

**Y Maharaj[1], S Parasnath[2]**

1. MBBCh,FCPath(SA)Haem, Subspecialty Trainee in Clinical Haematology, Department of Clinical Haematology, Inkosi Albert Luthuli Central Hospital, Durban, KwaZulu Natal, South Africa.
2. MBChB, FCPath(SA)Haem, Cert Clin Haem(SA). Head Clinical Unit Clinical Haematology, Department of Clinical Haematology, Inkosi Albert Luthuli Central Hospital, Durban, KwaZulu Natal, South Africa. University of KwaZulu Natal, Durban, KwaZulu Natal, South Africa.

## INTRODUCTION:

Chronic Lymphocytic Leukaemia (CLL) is an acquired clonal lymphoproliferative disorder leading to an accumulation of mature B lymphocytes which are functionally incompetent. CLL is relatively rare in South Africa compared to the western world, however it appears to have an aggressive clinical course in the black South African population and presents at a younger age.

## CASE REPORT:

- A 39-year-old black South African male with a background of HIV and hepatitis B co-infections on anti-viral treatment and virologically suppressed, presents with a 4-month history of significant bilateral lymphadenopathy and constitutional symptoms. Physical examination revealed a massive splenomegaly. An excision biopsy confirmed the diagnosis of CLL. Molecular testing demonstrated a 17p deletion in 85% of cells as the sole abnormality. Routine blood analysis, flow cytometry, bone marrow biopsy and a staging CT scan were performed as part of the work up. He was classified as Rai stage IV.
  Full blood count (FBC) revealed a leukocytosis of 68.07 x $10^9$/L, a severe anaemia of 4.9 g/dL, a thrombocytopenia of 29 x $10^9$/L and an absolute lymphocyte count of 64.67 x $10^9$/L. Although a direct anti-globulin test was not performed at the time, it was suspected that the anaemia may have been immune-mediated. The flow cytometry revealed a population of cells which expressed CD5+, CD19+, CD23+, dim CD20+, CD43+, CD79b-, FMC7- and surface lambda light chain restriction in keeping with a CLL phenotype. Symptomatic anaemia and sepsis complicated his multiple admissions.
  The treatment options in our setting are limited due to economic constraints. We elected not to treat him with FCR (fludarabine, cyclophosphamide and rituximab) due to severe and recurrent infections, as well as his poor performance status. We opted commence him on R-CVP (rituximab, cyclophosphamide, vincristine and prednisone) in an attempt to reduce the disease burden. We concurrently provided support with blood products and antibiotics. He has remained transfusion dependent. The suspected auto-immune haemolytic anaemia did not respond to high dose prednisone therapy. Intravenous immunoglobulin was administered in view of the ongoing infections.

## DISCUSSION:

Over the last few decades CLL therapy has undergone many changes. The treatment landscape of CLL in South Africa remains challenging due to the limited access to effective oral targeted therapies such as Bruton's Tyrosine Kinase (BTK) inhibitors and venetoclax, in the state-funded healthcare system. Selecting the correct treatment for the patient requires consideration of the patient's co-morbidities, disease characteristics and prior therapies. Patients with TP53 disruptions have a shorter progression free survival and overall survival when treated with standard immunochemotherapy regimens such as FCR. In view of the 17p deletion the ideal upfront therapy would be either a BTK inhibitor alone or venetoclax plus an anti-CD20 monoclonal antibody. We are hoping to start zanubrutinib pending a special motivation on a compassionate use programme. Zanubrutinib is a second generation covalent BTK inhibitor with higher specificity and less off-target inhibition than ibrutinib.

## CONCLUSION:

In conclusion we report a case of CLL in a young black man with a 17p deletion. This case demonstrates the importance of having access to novel therapies and further highlights the difficulties we have in treating CLL in a resource limited setting.

UNIVERSITY OF KWAZULU-NATAL ™
INYUVESI YAKWAZULU-NATALI

KWAZULU-NATAL PROVINCE
HEALTH
REPUBLIC OF SOUTH AFRICA