

Incidence of haematological malignancies, Eastern Cape Province; South Africa, 2004–2013

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ABSTRACT

Background: The incidence of haematological malignancies in Africa's rapidly urbanising populations is insufficiently explored. Reliable population-based cancer statistics, however, continues to be a scarce resource in Africa and tends to be urban biased with limited rural coverage. In addition, many haematological malignancies are regarded as rare cancers, a sub-group that often affects the young disproportionately and require advanced diagnostic services and facilities able to deliver costly sophisticated treatments. This study provides a first attempt to estimate the incidence of haematological malignancies among the Eastern Cape Province population of South Africa.

Method: Multiple public- and private sector data archives and resources were utilised to optimise the identification of incident cases, including clinical records; bone marrow; cytology; histology; flow cytometry and cytogenetic records. Crude incidence, age- and gender-standardised rates are presented and comparison made with existing national data and select data from other economically developed countries and global institutions.

Results: A total of 3603 incident cases were identified between 2004 and 2013. Mature lymphoid malignancies accounted for approximately 60% (n = 2153), myeloma/plasma cell neoplasms 13% (n = 465), acute leukaemia 17% (n = 596), chronic myeloid leukaemia 4% (n = 155) and other myeloproliferative neoplasms 6% (n = 234) when stratified according to conventional groups. Most subtypes increase with age, with male excess.

Conclusion: Haematological malignancies in the Eastern Cape Province show disparities in gender and pathology-specific incidence patterns. The present study suggest that haematological malignancies are not uncommon in this region and the incidence rate of at least one rare subtype, APL, is comparable with some European populations.

1. Introduction

Haematological malignancies (HMs) have been described in Africa and Southern Africa for over a century [1–4]. Early references to cancer from the previous century bear witness to longstanding and widespread interest in cancer prevalence. The narrative provides clues to changing patterns in cancer incidence. Some changes have been exposed as artefacts relating to improvements in diagnostic and registration practices. Some deviations represent real changes, for example the surge in lymphomas in the era of the HIV/AIDS pandemic [5]. The African continent, however, remains under-represented in global cancer statistics, with urban bias, limited rural coverage and limited reliable

population-based incidence data [6].

South Africa is described by the United Nations as a middle-income country, with indigenous populations akin to those in other developing countries urbanising at a fast pace [7]. The Eastern Cape Province (ECP) is regarded as one of the least urbanised regions, with vast and sparsely populated rural areas and is known for its large number of remaining traditional dwellings [8]. Although ECP has two highly developed metropolitan areas, the rural population continues to engage in traditional lifestyles, some through choice, but many through necessity imposed by widespread poverty and under-development [8].

Cancer surveillance in ECP entered the local and international arena in 1955 when high prevalence was detected for oesophageal cancer.

Abbreviations: ALL, acute lymphoblastic leukaemia; AML, acute myeloid leukaemia; APL, acute promyelocytic leukaemia; ASR, age standardised incidence rates; CI, confidence interval; CML, chronic myeloid leukaemia; ECP, Eastern Cape Province; HMs, haematological malignancies; HL, hodgkin lymphoma; ICD-O-3, International Classification of Disease for Oncology; IR, incidence rate; LPD, lymphoproliferative disorders; MGUS, monoclonal gammopathy of undetermined significance; MLUS, monoclonal B-cell lymphocytosis; NHL, non-hodgkin lymphoma; NPCR, National Program for Cancer Registries; RARECARE, surveillance of rare cancers in Europe

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The area soon gained the unenviable reputation as a “hotspot”. Several researchers contributed to this body of work [9–11]. A contemporary analysis of cancer incidence trends 1998–2012, showed that esophageal cancer remains the most common cancer for males [12]. The impact of HIV/AIDS has been significant in the region, with life-expectancy decreasing between 2000 and 2005 to 48.3 years [13]. HIV/AIDS and Tuberculosis (TB) accounted for 36,7% of deaths between 1997 and 2010 and cancers 7.4% [13].

In addition to esophageal cancer, HMs likewise have a history of provoking concern in this region. A scientific publication emerged in 1987 describing nine children who presented to the Red Cross War Memorial Children’s Hospital with a rare leukaemia subtype, acute promyelocytic leukaemia (APL) [14]. The authors expressed concern that cases presented to them within a relatively short period and that seven of the children originated from a “confined geographic space” within the Eastern Cape region [14]. No formal epidemiological study followed.

Historically there has been limited regional information available to clarify the burden of HMs in South Africa. The South African National Cancer Registry (NCR-SA) recently published the first incidence estimates for the country as a whole based mainly on histological diagnoses and with incomplete data contributions from the private sector [15]. The current study represents an attempt to produce tentative estimates of the incidence of HMs in the ECP, utilising all available public- and private sector laboratory archives and supplemented with relevant medical record data.

2. Materials and methods

2.1. Geographical location and population

ECP is located along the southeastern seaboard of South Africa. It is the second largest province in the country, the third most populous area after KwaZulu Natal and Gauteng, home to 14.4% of the South African population, with Black isiXhosa speaking South Africans comprising the majority [16]. According to the 2001 census, population density was estimated at 38 persons per km² (just above the national average of 37), with a slight increase to 39 per km² in 2011 [8,16]. The population can be represented as a broad based pyramid, typical of a developing country and with substantial out-migration of adults, notably males of working age (Fig. 1). This study utilised averaged mid-year census estimates for 2008 and 2009 as the denominator population to represent population exposed to risk between 2004 and 2013 [17].

2.2. Case finding

ECP meets its public health diagnostic laboratory requirements through a network of laboratories provided by the National Health Laboratory Service [18]. The present study collected data from regional public laboratory archives 2004 to the end of 2013 and from their private laboratory counterparts. Information specialists assisted to extract test-method codes and episode numbers registered for bone marrow examinations (cytology/histology), cytogenetics, tissue biopsy/histology and flow cytometry reports generated within and referred from ECP for the period of interest. All test episode numbers were manually screened to identify incident cases, confirm age and gender, exclude duplications and coding errors, confirm residence in ECP and correlate with cases previously captured in a pre-existing regional laboratory register. Laboratory resources were supplemented with information abstracted from medical records for new patients (2004 to 2013) registered at the main regional haematology unit. Institutional and ethics clearance were obtained from the Nelson Mandela University, the Department of Health, the CEO of the Port Elizabeth Hospital Complex, the NHLS, the University of Cape Town and also from PathCare and Ampath private laboratories.

2.3. Data analysis

Incident cases were abstracted for demographics (age, gender, area of residence), relevant clinical and laboratory information and diagnosis. The recommendations of the 2008 World Health Organisation (WHO) Classification of Tumours of Haematopoietic and Lymphoid Tissues were followed, along with the WHO’s International Classification of Diseases for Oncology-3 and ICD10 topography codes to categorise neoplasms according to lymphoid and myeloid lineages and identify relevant subtypes (see Table 1 for details on ICD 10 categories utilised for subtypes) [19,20]. Cases of monoclonal B-cell lymphocytosis (MLUS), monoclonal gammopathy of undetermined significance (MGUS) and myelodysplastic syndrome were not included in the present study. All data were de-identified and aggregated for statistical analysis. Cases that could not be assigned to a gender or age group or those of ambiguous lineage were excluded from gender, age and lineage specific analysis.

Descriptive demographics, age and gender specific annual crude numbers and annual crude incidence rates (IR) with 95% confidence intervals (CI) were computed according to 16 quinquennial groups commencing from 0 to 4 years and truncated at 75 years and above for

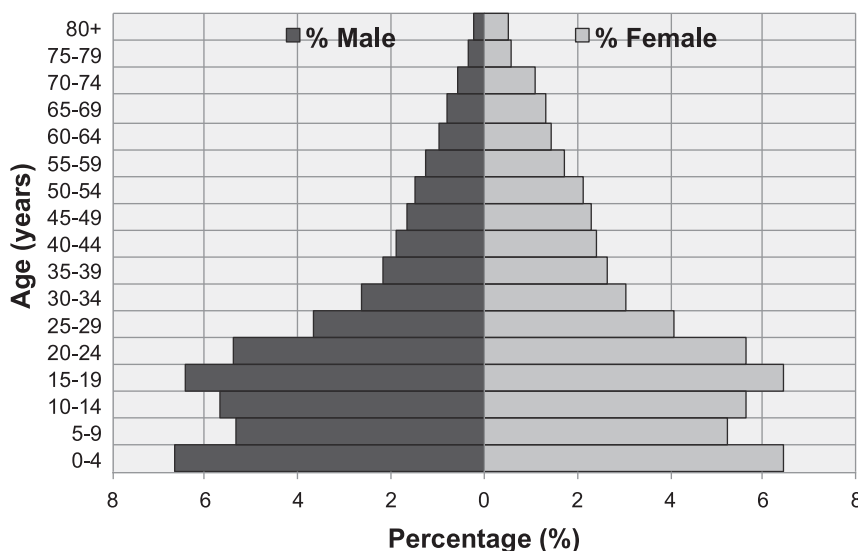


Fig. 1. Population pyramid, 2008/2009 mid-year averages, Eastern Cape Province, South Africa.

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