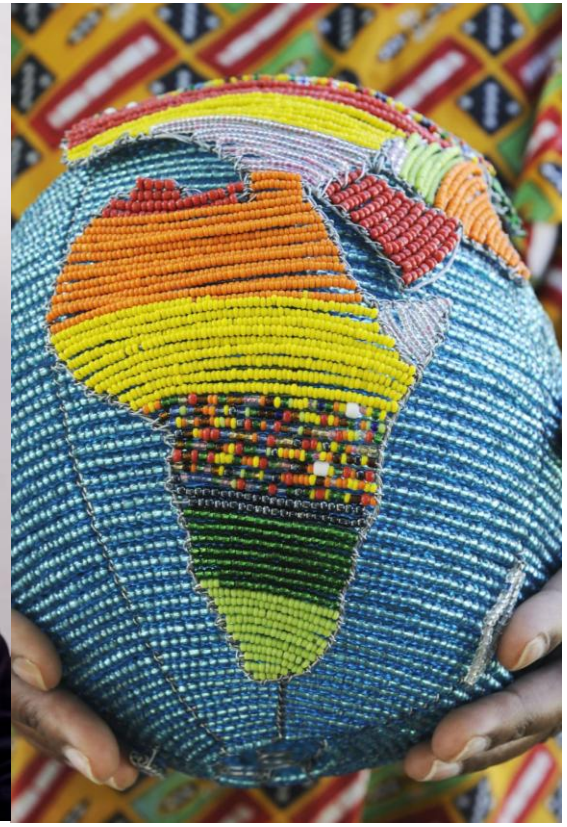
Retinoblastoma(RB) in Africa





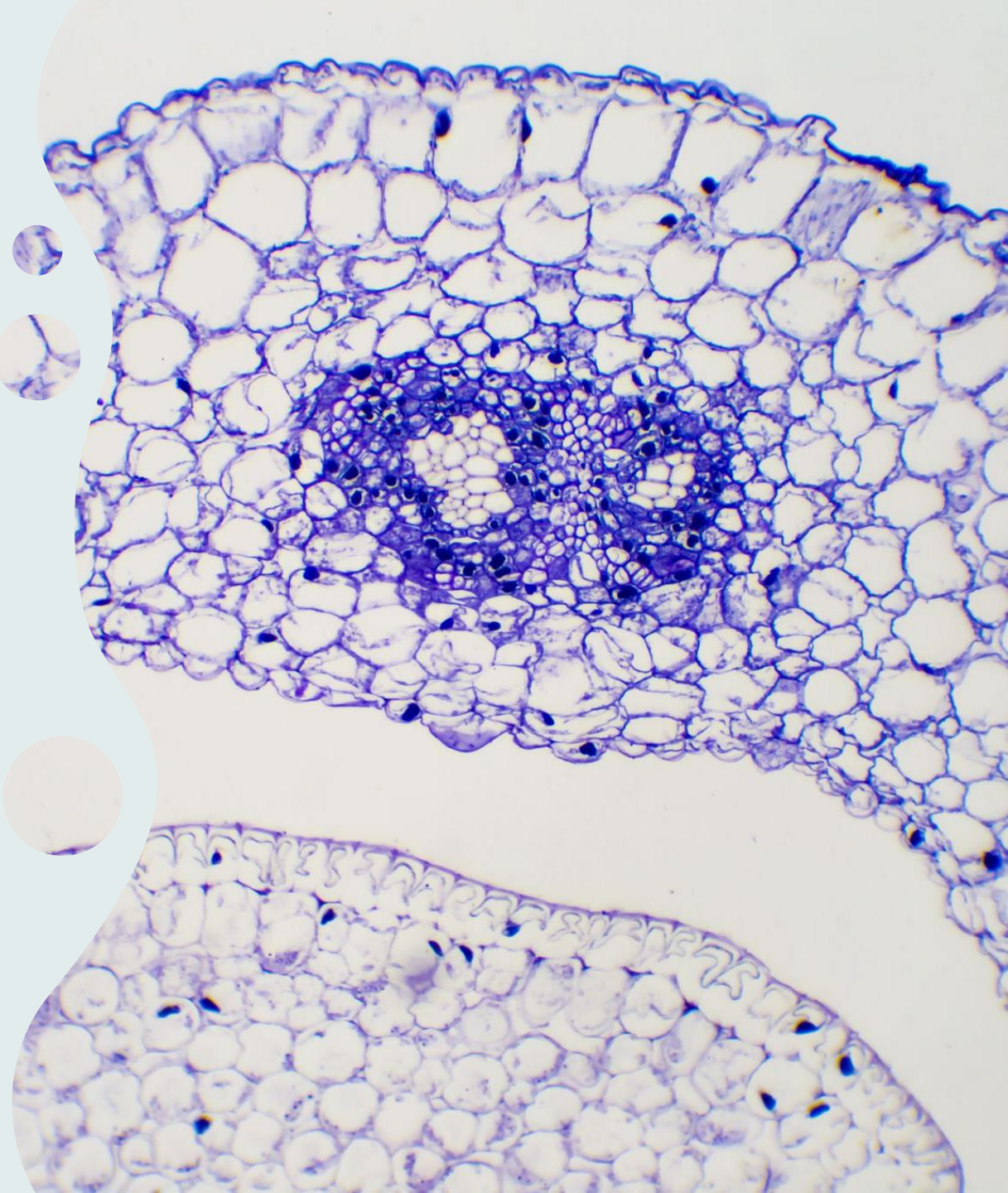
Dr. Pius Mwanja

Paediatric Ophthalmologist

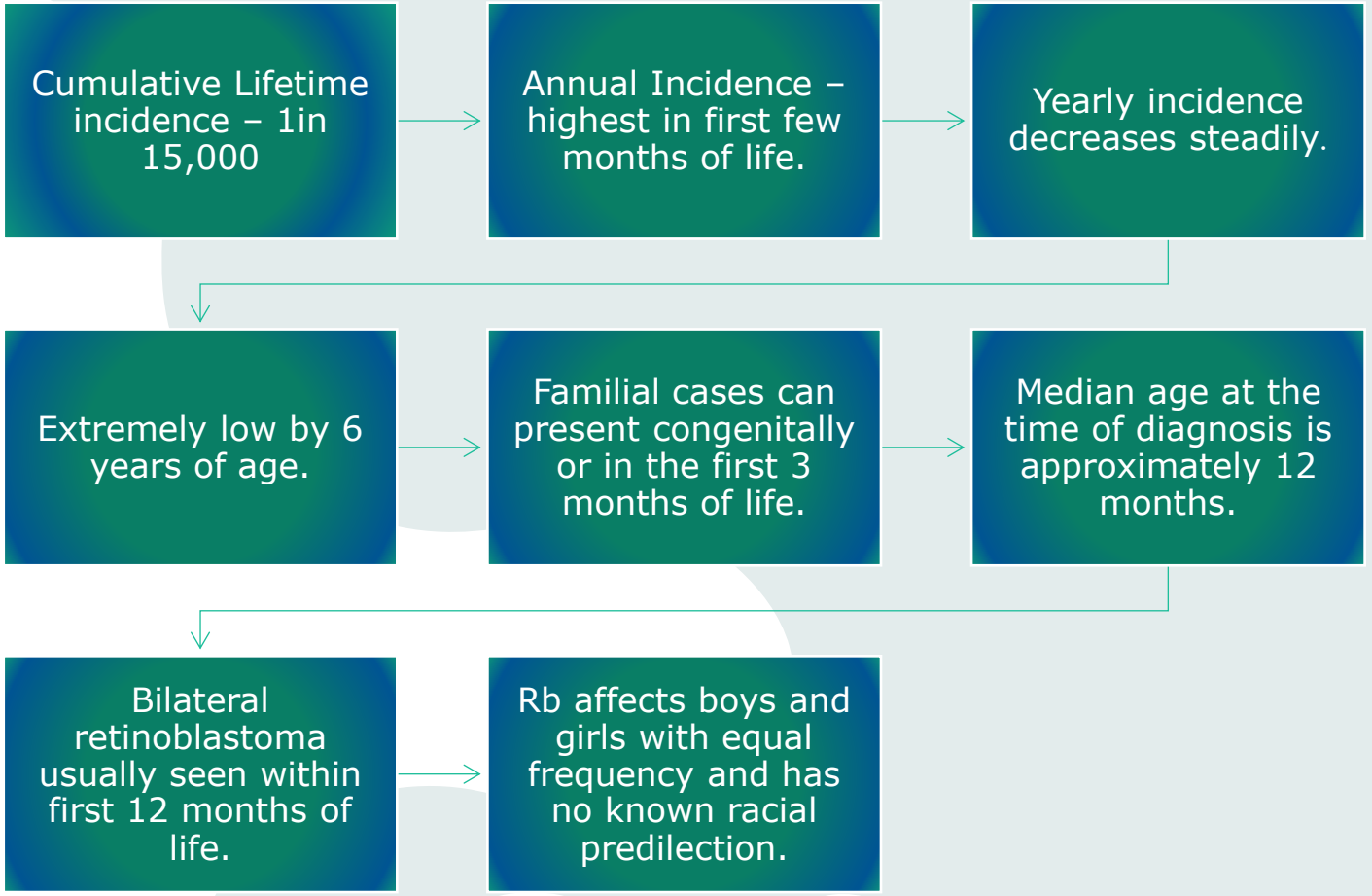
Ruharo Mission Hospital - Uganda

Introduction

- Primary malignant neoplasm of the retina that arises from immature retinal cells.
- It is the most common primary intraocular malignancy of childhood in all racial groups.
- Can be unifocal or multifocal.
- Unilateral (60 – 70%) or bilateral (30 – 40%).
- Sporadic (94%) or familial (6%)
- Non-hereditary (50-60%) or hereditary (40-50%)



General Epidemiology



Global Retinoblastoma Burden

RB is the most common pediatric ocular cancer and is typically curable through early diagnosis and protocol-based management. More than 95% of retinoblastoma cases occur before the age of 8 years. (1)

Globally, about 1 in 17,000 children develop retinoblastoma, an estimate of 7752 – 8914 newly- affected children each year. (2) (3) The incidence of retinoblastoma is known to vary with the size of the population and the birth rate. (4)

Tumor registers differ in countries. RB incidence in most low-income regions is estimated using mathematical methods based on population risk and live birth rates. Current estimates suggest that over half of RB cases originate in the Asia-Pacific region, with nearly a quarter from Africa. (5)

Sub-Saharan Africa and Uganda's Retinoblastoma burden

Most Retinoblastoma patients are from low- and middle-income countries (LMICs) with the highest incidence of retinoblastoma in the world in Africa and India. (3) (5)

Retinoblastoma (RB) is one of the most reported childhood cancers in Africa and was at some point the most common eye cancer overall in sub-Saharan Africa until overtaken by HIV-associated conjunctival carcinoma. (12)

Across a select 8 Sub-Saharan countries i.e., Ethiopia, Ghana, Malawi, Nigeria, Tanzania, Uganda, Zambia, and Zimbabwe, 1023 RB incident and familial cases are estimated each year. (6)

Compared with children in developing countries, the diagnosis of retinoblastoma in children in low-income countries is often delayed, resulting in children presenting with advanced retinoblastoma (i.e., combinations of proptosis, red eye, orbital cellulitis, and extraocular retinoblastoma) and thus poor patient survival. (3) (7) (8)

Mortality risk from retinoblastoma varies by economic status. In high-income countries, it's mainly due to second malignancies, while in low-income countries, most deaths occur from extraocular retinoblastoma and systemic metastasis. (9)

Uganda's Fight against RB

Uganda has a relatively large burden of children with retinoblastoma and statistically low levels of survival, but also, some health facilities like Ruharo Mission Hospital Eye Centre (REC) are working hard to improve Retinoblastoma care in the region. (10)

The East African region has only a few centers offering modern RB treatment. This scarcity has seen REC continue to serve the wider East African region as a tertiary RB treatment facility receiving patients from as far as South Sudan, DRC, and northern Tanzania among others (over 20% of our patient cohort are non-Ugandans)

REC, as a key ophthalmic hub and tertiary level Rb treatment centre in East Africa, experiences a significant number of new retinoblastoma cases per year (~120).

Retinoblastoma treatment at REC started in 2006 where the intervention was majorly surgical (Enucleation) with no chemotherapy. The introduction of neo-adjuvant chemotherapy 3 years later in 2009, in collaboration with St James Hospital Leeds, improved the survival of RB patients from 45% to 65%. (11)

Uganda's RB cancer treatment program will continue to rely significantly on awareness and prevention while it develops screening, diagnostic, and treatment capacities because of the limited resources available in the nation.

It is understandable that despite increased survival, late presentation is still the leading cause of death.

This emphasizes the significance of raising community disease awareness and screening, as well as accelerating accurate diagnosis and treatment of new cases by introducing much-needed advanced diagnostic equipment plus Low vision and rehabilitation/palliation for the survivors and advanced cases.

Various RB Clinical presentation patterns

A= leukocoria

B=secondary Glaucoma with Buphthalmos

C= red eye associated with uveitis

D= pseudo-Hypopyon

E= orbital inflammation

F= orbital invasion



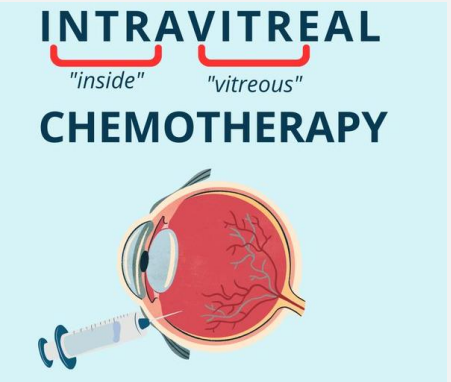
DIAGNOSTIC MODALITIES IN COUNTRY

MRI	YES	AVAILABLE IF PATIENT PAYS TO SOURCE IT FROM PRIVATE FACILITIES
CT Scan	YES	AVAILABLE IF PATIENT PAYS TO SOURCE IT FROM PRIVATE FACILITIES
ULTRASOUND	YES	
PATHOLOGY	YES	
GENETIC TESTS	NO	

Treatment modalities in country



- → **Intravenous chemotherapy: Yes**
- → **Intra-arterial chemotherapy: No**
- → **Intra-vitreal chemotherapy: Yes**
- → **Plaque brachytherapy: No**
- → **Laser therapy: Yes**
- → **Cryotherapy: Yes**
- → **External beam radiotherapy: Yes (at the Uganda Cancer Institute only)**



Conclusion

Developed nations demonstrate that retinoblastoma (RB) can be effectively cured with modern treatments. In contrast, Uganda like many African countries struggles with high mortality rates due to late diagnosis and limited treatment options.

Addressing these disparities is essential for improving outcomes for affected children.

References

1) Stefan DC: Patterns of distribution of childhood cancer in Africa. J Trop Pediatr 61:165-173,2015.

2) United Nations, Department of Economic and Social Affairs. World Population Prospects: the 2017 Revision. Volume I: Comprehensive Tables. https://population.un.org/wpp/Publications/Files/WPP2017_Volume-I_Comprehensive-Tables.pdf. Published 2017. Accessed January 18, 2020.

3) Fabian ID, Abdallah E, Abdullahi SU, et al. Global retinoblastoma presentation and analysis by national income level. JAMA Oncol 2020;6(5):685-695. doi:10.1001/jamaoncol.2019.6716

4) Usmanov RH, Kivelä T. Predicted trends in the incidence of retinoblastoma in the Asia-Pacific region. Asia Pac J Ophthalmol (Phila) 2014; 3: 151–57

5) Munier FL, Beck-Popovic M, Chantada GL, et al. Conservative management of retinoblastoma: challenging orthodoxy without compromising the state of metastatic grace. “Alive, with good vision and no comorbidity”. Prog Retin Eye Res 2019; 73: 100764

References

6) Hampejsková L, Bascaran C, Zondervan M. A tool for planning retinoblastoma services in sub-Saharan Africa. Pediatr Blood Cancer. 2017 Apr;64(4). doi: 10.1002/pbc.26296. Epub 2016 Nov 3. PMID: 27808454.

7) Dean M, Bendfeldt G, Lou H, et al: Increased incidence and disparity of diagnosis of retinoblastoma patients in Guatemala. Cancer Lett 351:59-63, 2014.

8) Tomar AS, Finger PT, Gallie B, et al. Global retinoblastoma treatment outcomes: association with national income level. Ophthalmology 2021;128: 740–53.

9) Chantada GL, Qaddoumi I, Canturk S, et al. Strategies to manage retinoblastoma in developing countries. Pediatr Blood Cancer 2011;56: 341–48

10) K. Waddell, M. Matua, C. Bidwell, R. Atwine, J. Onyango, S.V. Picton, I. Simmons, J. Stahlschmidt, W.T. Johnston, R. Newton, A ten-year study of Retinoblastoma in Uganda: An approach to improving outcome with limited resources, Cancer Epidemiology, Volume 71, Part B,2021,101777, ISSN 1877-7821,https://doi.org/10.1016/j.canep.2020.101777. (https://www.sciencedirect.com/science/article/pii/S1877782120301119)

References

11) Waddell KM, Kagame K, Ndamira A, Twinamasiko A, Picton SV, Simmons IG, Revill P, Johnston WT, Newton R. Improving survival of retinoblastoma in Uganda. Br J Ophthalmol. 2015 Jul;99(7):937-42. doi: 10.1136/bjophthalmol-2014-306206. Epub 2015 Jan 12. PMID: 25583281.

12) Kivelä T. The epidemiological challenge of the most frequent eye cancer: retinoblastoma, an issue of birth and death. Br J Ophthalmol. 2009 Sep;93(9):1129-31. doi: 10.1136/bjo.2008.150292. PMID: 19704035.



HAPPY NEW YEAR 2025