

Community Eye Health Journal



Early diagnosis and treatment can save the life of a child with retinoblastoma. **INDIA**

SWATHI KALIKI

Retinoblastoma: a curable, rare and deadly blinding disease



Richard Bowman
Honorary Clinical Consultant:
International Centre for Eye Health,
London School of Hygiene and Tropical
Medicine, London, UK.

Every year, thousands of babies and children in low- and middle-income countries lose their sight and their lives to a treatable childhood eye cancer called retinoblastoma; usually because it was not recognised and treated in time.

Although retinoblastoma is relatively uncommon, it can have devastating consequences for the children affected by it. If treated too late, it can lead to the loss of the eye, invasion of the brain and death.

Retinoblastoma does not affect everyone equally. In high-income countries, fewer than 5% of children die as a result of the condition, thanks to early diagnosis and specialist treatment. In Africa, however, it is typical to see 70% of children with retinoblastoma die, mainly because they presented too late. When mothers do present at a tertiary centre with a child who has advanced retinoblastoma, they often report that they have had several interactions with different health

professionals over many months or even years, but did not get the referral or care they needed. Every health professional reading this issue of the *Community Eye Health Journal* has a chance to redress this balance. We need to find and treat children with retinoblastoma early, before it causes disfigurement or death.

Doing so successfully requires adopting a multidisciplinary, multi-level and internationally collaborative approach that looks at the health system as a whole (see page 4). Raising awareness of retinoblastoma in the community, improving the detection and diagnosis of the condition, setting up good referral systems and offering good

Continues overleaf ►

© The author/s and *Community Eye Health Journal* 2018. This is an Open Access article distributed under the Creative Commons Attribution Non-Commercial License.



About this issue

Retinoblastoma is a rare condition with devastating consequences. If left untreated, it can lead to loss of the eye, invasion of the brain and death. In this issue, we offer information and guidance about the detection, diagnosis and treatment of retinoblastoma, including advice about screening for family members when genetic testing is not available, and a step-by-step guide to enucleation. We hope that every health professional reading this journal will raise awareness of this condition so that – globally – we have a better chance to save the life, eyes and sight of children with retinoblastoma.

- 1 Retinoblastoma: a curable, rare and deadly blinding disease**
Richard Bowman
- 5 A national retinoblastoma network: experiences in Kenya and the UK**
Ashwin Reddy and Kahaki Kimani
- 7 Understanding retinoblastoma: epidemiology and genetics**
Ido Didi Fabian and Mandeep S Sagoo
- 8 Germline retinoblastoma: estimating risk and counselling the family**
Elisabeth Rosser and Mandeep S Sagoo
- 10 Detecting retinoblastoma**
Manoj V Parulekar
- 11 Classification and staging of retinoblastoma**
Ido Didi Fabian, Ashwin Reddy and Mandeep S Sagoo
- 14 Managing and treating intraocular retinoblastoma**
Ashwin Reddy, Mukesh Jain and Vikas Khetan
- 17 Standard reporting of high-risk histopathology features in retinoblastoma**
Caroline Thaug and Esin Kotiloglu Karaa
- 18 Management of retinoblastoma with extraocular tumour extension**
Swathi Kaliki and Vijay Anand Reddy Palkonda
- 20 How to do an enucleation for retinoblastoma**
Swathi Kaliki
- 23 CLINICAL SKILLS: Testing the red reflex**
Richard Bowman and Allen Foster
- 24 TRACHOMA: Making gains sustainable: partnering with WASH to stop the transmission of trachoma**
Leah Wohlgenuth, Helen Hamilton and Tim Jesudason
- 25 ONCHOCERCIASIS: River blindness: reducing the prevalence of clinical disease**
Charles Mackenzie, Martin Kollmann, Sabine Specht and Yao Sodhalon
- 26 Questions and answers on retinoblastoma**
- 27 Picture quiz**
- 27 Announcements and resources**
- 28 KEY MESSAGES**

counselling and high quality treatment (including good prosthetics) are all needed to increase the uptake of services and save lives.

Ministry of health

Ministries of health have the power to make dramatic improvements to the early detection and treatment of retinoblastoma. They can:

- Create public health campaigns to raise awareness that 'seeing something white' inside a child's eye is a medical emergency
- Include basic ocular history taking and eye examination techniques in the curriculum of community nurses
- Offer subsidised access to specialist treatment for children with this life-threatening condition.

Parents must be made aware that they should seek help urgently if they see something white inside their child's eyes. Emphasise that parents should not let their child be turned away and must not take 'no' for an answer if they feel there is something wrong.

A worthwhile investment

Investing resources in the early detection and referral of children with retinoblastoma has wider benefits in the fields of childhood blindness in low- and middle-income countries, as the same criteria (something white in the eye) will also help with the early detection of childhood cataract. Late presentation of childhood cataract is the leading cause of treatable blindness in children, and is entirely preventable if cataracts are detected and treated in time.

In the community

Nurses and health workers seeing children in the community can check children's eyes during routine immunisation appointments, for example. **Something abnormal, white or shiny, or a squint, may be the first sign of retinoblastoma and requires urgent specialist referral.** Listen to

Community Eye Health Journal
VOLUME 31 • NUMBER 101 • 2018



Editor
Elmien Wolvaardt Ellison
editor@cejournal.org

Consulting editor for Issue 101
Allen Foster

Editorial administrator
Anita Shah
anita.shah@lshtm.ac.uk

Editorial committee
Nick Astbury
Matthew Burton
Sally Crook
Allen Foster
Clare Gilbert
Hannah Kuper
Fatima Kyari
Janet Marsden
Priya Morjaria
G V Murthy
Daksha Patel
Noela Prasad

Babar Qureshi
Serge Resnikoff
Richard Wormald
David Yorston

Regional consultants
Hannah Faal (AFR)
Kovin Naidoo (AFR)
Wanjiku Mathenge (AFR)
Van Lansingh (AMR)
Andrea Zin (AMR)
Ian Murdoch (EUR)
Janos Nemeth (EUR)
GVS Murthy (SEAR)
R Thulsiraj (SEAR)
Babar Qureshi (EMR)
Mansur Rabiou (EMR)
Leshan Tan (WPR)
Hugh Taylor (WPR)

Proofreader Caroline Thaug
Design Lance Bellers
Printing Newman Thomson

CEHJ online

Visit the *Community Eye Health Journal* online. All back issues are available as HTML and PDF. Visit: www.cehjournal.org

Online edition and newsletter
web@cehjournal.org

Please support us

We rely on donations / subscriptions from charities and generous individuals to carry out our work. We need your help.

Subscriptions in high-income countries cost UK £100 per annum. Contact Anita Shah admin@cehjournal.org or visit our website: www.cehjournal.org/donate

Subscriptions

Readers in low- and middle-income countries receive the journal free of charge. Send your name, occupation, and postal address to the address below. French, Spanish, and Chinese editions are available. To subscribe online, visit www.cehjournal.org/subscribe

Retinoblastoma

When should we suspect?



White reflex



Deviated eye



Red eye



Enlarged eye



Deviated eye with white reflex

If you see any of the above features,
see your doctor immediately!



Shrunken eye

Recognise the
disease early and
save a child's life

the parents and/or carers. If they have seen something white or abnormal in their child's eye, believe what they say, take it seriously and seek specialist advice.

In Tanzania, community nurses have been trained to examine the red reflex (p. 23) using an Arclight ophthalmoscope. The Arclight is an affordable, solar-powered and easy-to-use ophthalmoscope. It has shown preliminary promise; the community nurses found it easy to learn and began picking up cases of

cataract and retinoblastoma by using it. Nurses can learn how to examine the red reflex at the same time as examining the child's other systems.

Tertiary centres

At tertiary centres, histopathologists have a crucial role: once the eye is removed the child may be able to leave hospital completely cured or may need chemotherapy

Continues overleaf ►

Public health awareness campaigns can support the early detection of retinoblastoma
INDIA

Address for subscriptions

Anita Shah, International Centre for Eye Health, London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK.

Tel +44 (0)207 958 8336

Email admin@cehjournal.org

Correspondence articles

We accept submissions of 800 words about readers' experiences.

Contact Anita Shah:

correspondence@cehjournal.org

Published by the International Centre for Eye Health, London School of Hygiene & Tropical Medicine.

Unless otherwise stated, authors share copyright for articles with the *Community Eye Health Journal*. Illustrators and photographers retain copyright for images published in the journal.

Please note that articles are published online first and may have been shortened to fit the available space in this printed edition.

Unless otherwise stated, journal content is licensed under a Creative Commons Attribution-NonCommercial (CC BY-NC) license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial purposes, provided that the copyright holders are acknowledged.

ISSN 0953-6833.

Disclaimer

Signed articles are the responsibility of the named authors alone and do not necessarily reflect the views of the London School of Hygiene & Tropical Medicine (the School). Although every effort is made to ensure accuracy, the School does not warrant that the information contained in this publication is complete and correct and shall not be liable for any damages incurred as a result of its use.

The mention of specific companies or of certain manufacturers' products does not imply that they are endorsed or recommended by the School in preference to others of a similar nature that are not mentioned. The School does not endorse or recommend products or services for which you may view advertisements in this Journal.

The *Community Eye Health Journal* is supported by:



or radiotherapy; this decision must be based on accurate histopathological staging (page 18).

International collaboration

To promote this multi-level, multi-disciplinary and internationally collaborative approach, the Commonwealth Eye Health Consortium has provided start-up funding for an Rb-Network known as Rb-NET, which has already generated specific country plans, a set of core outcome indicators, best practice protocols and a practical resource manual (<http://cehc.lshtm.ac.uk/dr-links/rbnet/>).

Basic clinical research questions still need to be answered. For instance, researchers in Uganda have shown an improvement in survival by giving chemotherapy before surgery on the basis that so many children have extra-ocular spread at time of presentation. On the other hand, a small study from Tanzania showed that 60% of children for whom there was good histology after enucleation had complete excision of the tumour with low risk and never needed chemotherapy. So which should come first in these settings – chemotherapy or surgery? By combining multi-centre and multi-country clinical research, as Rb-NET has started to do, we can begin to answer these questions and prevent needless tragedies.

This issue of the *Journal* demonstrates that there is real momentum and determination to improve outcomes for children with Rb in all countries across the world. It contains concise, practical information that should help all of us to make a difference.

I am a child, not a case



Abby White
Chief Executive: World Eye Cancer Hope UK
www.wechope.org

Patients with retinoblastoma are first and foremost children who happen to have cancer in their eye(s). Defining a child as a ‘case’ dehumanises them and draws attention away from thoughts about their complete wellbeing and that of their family.

A child with retinoblastoma is not a medical specimen. They are a complete individual with thoughts, feelings, hopes, dreams, likes and dislikes. They have the ability to generate every kind of emotion in those who care for them. They are desperately loved, and most parents would give their own eye if it could spare their child’s suffering.

In evaluating different treatments, we must weigh the value of each treatment in relation to the child’s complete wellbeing. We must look beyond the physical body to embrace and care for the child’s emotional health, during therapy and long after into adulthood.

Perhaps if we collectively take care to consider ‘children’, ‘families’ and ‘survivors’ rather than ‘cases’, we will together establish a level ground on which we can both treat the cancer, and heal the spirit in equal measure to set the child up for a healthy, happy future.

Rb-NET website

<http://cehc.lshtm.ac.uk/dr-links/rbnet/>

Resource manual

<http://cehc.lshtm.ac.uk/resource-manual-for-rb-management-v-sept-1-002/>

Arclight

www.arclightscope.com

Table 1 Roles and responsibilities in the detection, referral and treatment of retinoblastoma

Individual responsibilities			
Parent →	Health worker/nurse →	Ophthalmologist →	Specialist eye centre
<p>Seek help urgently if you see something white inside the centre of the eye (the pupil) OR if you take a photograph and only one eye has a red dot in the centre</p> <p>Do not let anyone turn you away and do not take no for an answer until a doctor at a hospital has examined the child’s eyes using a bright light</p>	<p>Believe the parents if they say they have seen something white inside the pupil and seek specialist advice. Treat it as a medical emergency</p> <p>Learn how to test the red reflex (p. 23). Test all children during routine visits and immunisations</p>	<p>Learn to recognise retinoblastoma and to identify eyes that need enucleation</p> <p>Counsel parents about the good cosmetic outcomes of enucleation with implantation. Show pictures of children with good outcomes</p> <p>Learn how to enucleate, taking more than 15 mm of optic nerve. Always examine the fundus of the fellow eye when you perform an enucleation: there could be a small tumour which is treatable by laser</p> <p>Refer all children with signs of retinoblastoma in two eyes to a national or specialist centre for urgent treatment</p>	<p>Same as for ophthalmologists, plus:</p> <p>Learn how to give focal or laser treatment to smaller tumours (usually in the second eye)</p> <p>Create multidisciplinary teams who work closely together to coordinate the treatment of each child</p> <p>Include in this team: ophthalmologists, oncologists, histopathologists, nurses, child life specialists or play therapists and/or counsellors</p> <p>Offer general and genetic counselling to parents/carers</p> <p>Refer parents to other sources of support for their child’s learning and development</p>
The Ministry of Health’s responsibilities towards the above			
<p>Run public awareness campaigns so that parents know that treatment is possible and know when to see a doctor</p>	<p>Ensure that the red reflex test (p. 23) is included in the curriculum for nurses and health workers</p>	<p>Ensure there is at least one ophthalmologist per 100,000 population</p>	<p>Support the development of national retinoblastoma centres and referral networks. Offer subsidised access to specialist treatment for all children with retinoblastoma. Provide screening services for siblings and accommodation or travel subsidies for the parents or carers of these young children.</p>