

# Late-stage retinoblastoma in older children: Case-based lessons and a call for early diagnosis in resource-limited settings

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## Abstract

**Background:** Retinoblastoma (RB) typically presents before age five; presentation beyond this age is uncommon but documented. We describe a 9-year-old Nigerian girl who presented with a fungating orbital mass secondary to advanced unilateral RB. The child had an 11-month history of progressive leukocoria and proptosis initially misinterpreted as an infection, followed by multiple delays at the primary-care level and prolonged use of traditional remedies. Factors contributing to the delay in diagnosis and timely presentation for specialist care included poor awareness of early RB signs, misdiagnosis, delayed referral, and sociocultural practices.

**Main discussion:** The case underscores how limited community awareness, mismanagement at the primary-care level, and reliance on traditional remedies prolong the diagnostic timeline until curative treatment is less feasible. These case-specific delays illustrate broader systemic gaps in early detection and referral in LMICs.

**Conclusion:** While advanced treatment and genetic tools hold promise, survival for RB in LMICs still depends on timely diagnosis and prompt intervention. This case highlights the urgent need for improved early recognition and referral systems, while acknowledging that late-age diagnosis has also been reported globally. Bridging the awareness gap and investing in sustainable early detection strategies must be prioritized to close the survival divide between high- and low-income settings.

**Keywords:** Retinoblastoma, Late presentation, Pediatric cancer, Early diagnosis, Low-resource settings, Ocular oncology

## Introduction

Retinoblastoma (RB) is one of the success stories of pediatric oncology when detected early, with survival rates approaching 99% in high-income settings where prompt recognition, advanced imaging, and modern treatment options are readily available [1–4]. In stark contrast, children in many low- and middle-income countries (LMICs) continue to face delayed diagnosis, advanced disease at presentation, and unacceptably high mortality rates [5,6]. The recently reported case of a 9-year-old girl presenting with fungating RB at the Evangelical Church Winning All (ECWA) Eye Hospital, Nigeria, painfully illustrates how systemic barriers and delays can turn a highly curable disease into a cause of avoidable death [6].

Although delayed diagnosis is more frequently associated with LMICs due to system-level barriers, studies from high-income settings have also documented retinoblastoma presentation beyond the typical early-childhood age window. Clinical and genetic characteristics of children diagnosed after

the age of five, some of whom presented without the usual early symptoms, have been reported [7]. Similarly, cases occurring in children older than six years have been documented, demonstrating that late-onset or late-diagnosed RB, while uncommon, is not restricted to resource-limited environments [8]. These findings highlight that although LMICs bear a disproportionately heavy burden of late presentation, delayed diagnosis can occasionally occur across different healthcare settings.

Available data from sub-Saharan Africa indicate that approximately 40–50% of children with RB still present with extraocular disease at first diagnosis, compared with less than 10% in high-income countries [6]. Smaller national studies from Nigeria consistently report even higher rates, ranging from 60% to over 70% [1,9,10], underscoring the urgent need for strengthened early detection strategies. These statistics confirm that the present case is not an isolated occurrence but reflects a persistent pattern of delayed diagnosis and poor outcomes in many resource-limited settings.

Several studies have reported important patterns that support the relevance of this communication. Analyses show that delayed diagnosis remains more frequent in LMICs, with a higher proportion of extraocular disease at presentation and greater mortality risk. At the same time, studies from higher-income settings have documented that retinoblastoma can also be diagnosed after the age of five or six years, even though this is less common [7,8]. These findings highlight both shared and context-specific challenges and underscore the need to examine the systemic factors that contributed to the late diagnosis in this case.

This rapid communication adds to existing literature by illustrating how a combination of caregiver factors, primary-care missteps, and sociocultural practices can delay diagnosis far beyond the typical age range. By analyzing this case within the broader global evidence, we highlight context-specific gaps in early detection and offer practical approaches to reduce late presentations in similar settings.

### **Insight from the Report**

In the published case [6], a young girl presented with unilateral retinoblastoma well beyond the typical age window for early diagnosis, which is usually by the age of three years. By the time of presentation, the disease had progressed to a fungating orbital mass with optic nerve involvement; clear evidence of advanced extraocular spread [11,12]. Despite prompt surgical exenteration and adjuvant chemotherapy, the patient's prognosis remained poor due to the extent of late-stage disease, and she sadly succumbed within two months of diagnosis. This case starkly demonstrates how late presentation not only increases mortality but also limits the effectiveness of otherwise available treatments. Such tragic outcomes underscore the persistent gaps in community awareness, early detection, and timely referral that continue to challenge retinoblastoma control in resource-limited settings.

The child had an 11-month history of progressive symptoms, beginning with redness, tearing, and visual decline. These were initially managed at a local clinic as presumed infection with topical antibiotics and corticosteroids, without imaging or specialist referral. As symptoms progressed to proptosis and increasing pain, the family sought help from traditional healers before eventually presenting to the tertiary center. Clinical examination revealed a large fungating mass with no light perception, while B-scan ultrasonography and

CT imaging confirmed vitreous infiltration, calcifications, and optic nerve involvement. She underwent modified exenteration followed by two cycles of vincristine, etoposide, and carboplatin. Although she tolerated treatment, she developed progressive systemic symptoms suggestive of metastatic disease and died during the second month of follow-up.

### **Contributing Factors**

A major contributing factor to this delayed presentation is the limited awareness among caregivers regarding the early warning signs of RB, such as leukocoria or strabismus [1,6,13]. In many communities, parents may misinterpret a white pupil reflex or visual changes as minor or temporary problems, and they may lack access to trusted health information sources that could guide them toward timely care-seeking behavior. Often, these early signs are overlooked until more severe symptoms such as proptosis, pain, or ulceration force families to seek help by which time the disease is usually advanced.

Equally important is the role of the primary care level in either facilitating or hindering early detection. Evidence shows that in many low-resource settings, frontline health workers frequently misinterpret the early signs of retinoblastoma as common eye infections or inflammatory conditions, resulting in inappropriate prescriptions for antibiotics or corticosteroids instead of urgent referral to specialist care [5,14,15]. This pattern significantly contributes to diagnostic delays and the high proportion of children presenting with advanced disease. The lack of standardized referral protocols, combined with training gaps and limited diagnostic capacity in peripheral health facilities, means that valuable time is often lost during the critical early stages; when timely enucleation or focal therapies could still be curative.

Cultural beliefs and practices can further compound these challenges. In many communities, families may initially seek help from traditional healers or spiritual leaders for alternative remedies before turning to formal medical services [16,17]. These detours often delay evidence-based treatment and allow the tumor to progress unchecked. Additionally, financial barriers, long travel distances to specialized centers, and fear of surgery can all contribute to prolonged delays in seeking definitive care [18]. In some families, the social stigma associated with removing an eye, especially when it leaves a child with only one eye can further discourage timely acceptance of life-saving treatment. This stigma often deepens parental hesitation and fuels a preference for alternative remedies or prolonged watchful waiting, which can allow the disease to advance beyond curable stages.

### **Opportunities for Improvement**

Addressing these multifaceted challenges requires coordinated, context-specific solutions that tackle the problem from multiple angles. One crucial step is to implement sustained community awareness campaigns that educate caregivers about the early warning signs of retinoblastoma and the importance of seeking prompt medical care [5,18]. Public health messaging delivered through radio broadcasts, community health outreach, and integration into routine maternal and child health visits can empower parents to recognize suspicious signs such as a white pupil reflex or eye misalignment and act without delay.

Strengthening the capacity of frontline healthcare workers is equally essential. Regular training workshops and the inclusion of RB “red flag” indicators in standard primary care guidelines can better equip nurses, community health extension workers, and general practitioners to recognize potential cases and refer them quickly [6,18]. Establishing clear, well-communicated referral pathways between local clinics and tertiary eye centers can further minimize the time lost between first presentation and definitive treatment.

While widespread genetic screening for RB remains impractical in many low-resource settings, targeted genetic counseling and surveillance for families with a known hereditary RB history can improve early detection among siblings and future generations. Building trust with affected families and demystifying genetic testing are important steps toward making this approach acceptable and accessible.

Long-term progress also depends on robust investment in local oncology infrastructure [1,5]. International partnerships and capacity-building initiatives can help equip hospitals with modern diagnostic tools, advanced treatment options, and strong multidisciplinary teams capable of delivering timely, effective care. Mentorship arrangements, collaborative research, and training exchanges with established ocular oncology centers can help local hospitals grow into regional hubs of expertise.

Encouragingly, pilot programs such as the Kenyan National Retinoblastoma Strategy and Nigeria’s Vision 2020 initiatives have demonstrated that community-based screening and the integration of ocular oncology into routine child health services can significantly increase early detection rates [18–21]. Expanding and adapting these successful models to other regions could help bridge the gaps in awareness, timely diagnosis, and access to life-saving treatment that this case so clearly highlights.

### **Screening and Routine Eye Examination Practices**

In our practice, children are generally examined opportunistically during routine child-health visits or when they present with symptoms. Newborn checks may include basic assessments such as external eye inspection and red-reflex testing, although implementation varies across facilities. Guidance for early eye assessment is drawn from national child-health policies, WHO recommendations, and professional guidance from the Ophthalmological Society of Nigeria. However, many primary healthcare centers have limited equipment and training, which affects their ability to reliably detect early ocular abnormalities, including conditions such as retinoblastoma.

Early detection efforts often rely on integrating eye-health messages into maternal and child-health services, training primary-care workers to recognize warning signs, and encouraging timely referral to tertiary eye hospitals. Evidence shows that school-based eye-screening programs in Nigeria are inconsistent, and many school-aged children have never undergone an eye examination. For example, in a large southern-Nigeria survey, 89.9% of children had never had any previous eye check [22]. Similarly, a Lagos-based study found that many children with amblyopia remain undiagnosed due to the absence of routine school screening programs [23]. These gaps contribute to inconsistent early identification of childhood eye diseases including amblyopia, congenital cataracts, and retinoblastoma and help explain why delayed presentation remains common in many settings.

### **Broader Implications**

Across different regions of the world, improving outcomes for retinoblastoma requires attention to several shared priorities, including strengthening caregiver awareness, enhancing the ability of frontline health professionals to recognize early signs, building well-equipped multidisciplinary ocular oncology teams, providing financial and social support for families, and implementing measures that reduce treatment abandonment.

While delayed diagnosis is more frequently observed in low- and middle-income countries (LMICs) due to system-level barriers, late-age presentation has also been reported in higher-income settings. Clinical and genetic features of children diagnosed after the age of five have been documented [7], and cases occurring in children older than six years have likewise been described in another report [8]. These findings indicate that late-onset or late-diagnosed RB, although uncommon, is not exclusively a phenomenon of resource-limited environments and may occur across different healthcare settings, generally at lower frequency. Nonetheless, system-level barriers and socioeconomic factors in LMICs substantially increase the risk of advanced-stage disease at first presentation.

The pattern highlighted in this case mirrors trends widely reported in sub-Saharan Africa and parts of Asia, where retinoblastoma frequently presents at an advanced, extraocular stage, resulting in lower survival rates compared to high-income regions. While early detection remains the cornerstone of improving outcomes, ensuring continuity of care through structured, reliable follow-up systems is equally vital. Many families disengage from care after the initial phase of treatment due to prohibitive costs, travel challenges, or lack of community support, leading to preventable relapse and avoidable mortality.

Establishing robust patient tracking systems, supported by community health worker follow-up, can help families stay engaged throughout the treatment journey and monitor at-risk siblings when hereditary risk exists. Unlike high-resource settings, where the focus has long shifted toward vision preservation and rehabilitation, many children in low-income countries still lose their lives to this otherwise curable disease. Addressing these systemic gaps must remain a priority to close the survival divide and ensure every child with retinoblastoma has a fair chance at life and sight.

### **Conclusion**

The tragic outcome described in this case stands as a powerful reminder that retinoblastoma remains a preventable cause of childhood blindness and death when detected and treated in a timely manner. While advances in therapy and modern genetic tools hold great promise, they must be paired with grassroots strategies that address the underlying causes of delayed presentation and diagnosis. Closing the awareness gap, equipping primary healthcare providers with the skills to suspect and refer cases early, supporting families through counseling and education, and investing in resilient oncology services are all critical steps toward narrowing the survival divide.

It is imperative that governments, local health authorities, non-governmental organizations, and international partners work together to translate these lessons into clear policies, sustainable funding, and community-based programs. Only through collective, coordinated action can we ensure that no child loses their sight or their life to a preventable and treatable cancer like retinoblastoma.

## Authors' Contributions

All authors contributed equally, read and approved the final manuscript.

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## Competing Interest

There are no conflicts of interest.

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