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## Original article

# Multidisciplinary management of retinoblastoma: Experience in 37 eyes<sup>☆</sup>



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### ABSTRACT

**Objective:** To report the results of the multidisciplinary management of patients with retinoblastoma, including survival, enucleation rate, and systemic chemoreduction success. **Methods:** A retrospective study was conducted on 27 patients (37 eyes) diagnosed with retinoblastoma, and treated by a multidisciplinary team in San Juan de Dios Hospital. Demographic information, clinical characterization, survival, local and systemic treatments were included in the analysis. Patients treated with intra-arterial chemotherapy (IAC) were also reviewed.

**Results:** The study included 14 male patients (52%). The median of age at presentation was 8 months (0.16–90). The median follow-up time was 33 ± 21 months. The diagnosis was made in 10 (37%) cases after 15 months old, with a median of 35 months (24–90). 17 (63%) patients had unilateral retinoblastoma, and 10 (37%) bilateral retinoblastoma. Leukocoria, isolated or associated with other signs, was the most frequent reason for referral (63%). Global enucleation rate was 57% (n = 21), being the primary treatment in 15 (55%) patients. Enucleation rate in unilateral retinoblastoma was 76.5%, and for bilateral retinoblastoma, it was 60% for one eye and 10% for both. Systemic chemotherapy was prescribed in 17 (63%) patients, with a mean number of cycles of 5.3 ± 2.1. The overall success of chemoreduction and focal therapy in order to avoid external radiotherapy and/or enucleation was 68%. Three patients were treated with IAC as a salvage therapy, controlling the tumor in 2 patients at 6 months of follow-up. These are the first cases reported in Chile. Survival rate was 100%.

**Conclusion:** Multidisciplinary management of retinoblastoma led to a survival rate and morbidity comparable with international reports.

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## Manejo multidisciplinario en retinoblastoma: experiencia en 37 ojos

### R E S U M E N

#### Palabras clave:

Retinoblastoma  
Quimioterapia  
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Quimioterapia intraarterial

**Objetivos:** Reportar los resultados del manejo multidisciplinario de pacientes con retinoblastoma, incluyendo supervivencia global, tasa de enucleación y éxito de la quimiorreducción sistémica.

**Métodos:** Estudio retrospectivo 27 pacientes (37 ojos) con diagnóstico de retinoblastoma, tratados por un equipo multidisciplinario en el Hospital San Juan de Dios. Se incluyeron información demográfica, características clínicas, supervivencia, tratamiento local y sistémico. Se realizó además un subanálisis de tratamiento con quimioterapia intraarterial (QIA).

**Resultados:** Catorce pacientes (52%) fueron de sexo masculino. La mediana de edad al ingreso fue de 8 meses (0,16-90). La mediana  $\pm$  desviación estándar de seguimiento fue de  $33 \pm 21$  meses. Diez casos (37%) se diagnosticaron después de los 15 meses de edad, con una mediana de 35 meses (24-90). En 17 (63%) pacientes fueron retinoblastomas unilaterales, mientras 10 (37%) tuvieron retinoblastoma bilateral. La leucocoria, aislada o asociada a otros signos, fue el motivo de consulta más frecuente (63%). La tasa global de enucleación fue del 57% ( $n=21$ ), siendo el tratamiento primario en 15 (55%) pacientes. La tasa de enucleación en retinoblastoma unilateral fue del 76,5% y en retinoblastoma bilateral del 60% de un ojo y el 10% de ambos. Diecisiete (63%) pacientes recibieron quimioterapia sistémica (media de ciclos:  $5,3 \pm 2,1$ ). El éxito global de la quimiorreducción sistémica y la terapia focal fue del 68%. Tres pacientes fueron tratados con QIA como terapia de rescate, logrando controlar el tumor en 2 pacientes a 6 meses de seguimiento, siendo los primeros casos en Chile. La supervivencia fue del 100%.

**Conclusión:** El manejo multidisciplinario del retinoblastoma permite una supervivencia y una morbilidad comparable con la literatura internacional.

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## Introduction

Retinoblastoma is a curable ocular cancer which, if not treated, could cause death within 2 years.<sup>1</sup> Its incidence is close to 1 for every 16,000 live births,<sup>2</sup> although it can reach one for every 34,000.<sup>3</sup> It is estimated that 13 new cases arise every year in Chile.<sup>4</sup> Most of these cases are cared for by the public system by means of the Explicit Health Guarantees (*Garantías Explícitas en Salud*, GES). The San Juan de Dios Hospital is a tertiary oncology referral center where these patients are treated by a multidisciplinary team.

The main goal of retinoblastoma management is patient survival, in addition to preserving the compromised eye and visual function as far as possible.<sup>5</sup> It involves a complex clinical process for discarding simulating lesions, detection of disseminated disease, selection of personalized therapy and strict follow-up for diagnosing recurrences. There is a range of treatments comprising systemic management with several modes of chemotherapy and focal ophthalmological treatments.

The primary objective of the present study is to report the result of multidisciplinary management of patients with retinoblastoma in our hospital, including overall survival, enucleation rate and success of systemic chemoreduction. The secondary goal is to report the characterization of

ophthalmological variables, the range of treatments in this series, including the first cases of intra-artery chemotherapy (IAC) carried out in Chile.

## Methods

Retrospective study of clinical histories which included patients diagnosed for retinoblastoma admitted to the San Juan de Dios Hospital between August 2007 and March 2014. The study excluded patients with retinoblastoma-simulating lesions (pseudo-retinoblastoma) and patients with retinoblastoma diagnostic but were in follow-up when the study began. The data collected comprised demographic data, reason for the visits, follow-up, family history, supplementary examinations, characterization of tumors, staging, classification,<sup>6,7</sup> systemic treatments (chemotherapy modes) and local treatments (including laser, cryotherapy, brachytherapy, IAC [topotecan 1 mg, melfalan 4 mg] enucleation), relapses and dissemination. The results of available biopsies were reviewed. The study was approved by the Ethics Committee of the institution. The genetic study was carried out in an international laboratory (Impact Genetics Inc., Canada). The statistical analysis was made with SPSS 16.0 2007 for Mac (SPSS Inc., Chicago, USA), utilizing *T* for Student and Chi square tests.

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