



# *kinderkrebsinfo*

*Informationsportal zu Krebserkrankungen bei Kindern und Jugendlichen*

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## **Stages of retinoblastoma (classification)**

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Author: Maria Yiallourous, created 2023/06/09, Release: Prof. Dr. med. Petra Ketteler, PD  
Dr. med. Eva Biewald, Prof. Dr. Dr. h.c. Nikolaos Bechrakis, Last modified: 2023/06/14

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## Table of Content

1. Classification of intraocular retinoblastoma (ICRB staging system) .....	3
2. Classification of extraocular retinoblastoma (IRSS staging system) .....	4
Bibliography .....	6



# Stages of retinoblastoma (classification)

Based on the extent, retinoblastoma is divided into different stages (also known as classification). Consideration is primarily focussed on whether the retinoblastoma is intraocular, thereby affecting the eye(s) only, or if the disease has spread to other parts of the body (extraocular retinoblastoma). Tumour extent is always assessed separately for both eyes.

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For staging of intra- and extraocular retinoblastoma, various classification systems have been developed; the currently most validated ones (ICRB and IRSS classification) will be outlined later. Another classification, the TNM classification for retinoblastoma, merges information on intra- and extraocular disease.

## 1. Classification of intraocular retinoblastoma (ICRB staging system)

Classification of intraocular retinoblastoma is mostly based on the so-called ABC classification. This International Classification of Retinoblastoma, (ICRB, Philadelphia version) considers size and site of the tumour as well as the existence and extent of spread within the vitreous body. This results in a classification of retinoblastoma into 5 stages (A to E).

**Stages of retinoblastoma according to the „International Classification of Retinoblastoma“ (ICRB, Philadelphia version) and treatment options**

Stages	Tumour characteristics	Therapy options
	Retinoblastoma with a maximum diameter of 3 mm <i>and</i> with sufficient distance to central structures such as the fovea centralis (area of highest visual acuity) and the blind spot (optic papilla, where the optic nerve leaves the orbit)	Laser coagulation Thermotherapy Cryotherapy Brachytherapy
	Retinoblastoma larger than 3 mm in diameter <i>or</i> distance between retinoblastoma and fovea centralis 3 mm <i>or</i> less distance between	Laser coagulation Thermotherapy Cryotherapy Brachytherapy Systemic chemotherapy Intra-arterial chemotherapy



Stages	Tumour characteristics	Therapy options
	retinoblastoma and blind spot 1.5 mm or less detachment of the retina due to tumour-induced liquid accumulation beneath the retina (maximum of 3 mm measured from tumour margin)	
	Tumours with defined margins and minimal local spread (underneath the retina and/or towards the vitreous body)	Systemic chemotherapy Intra-arterial chemotherapy Intravitreal chemotherapy (followed by consolidation)
	Large tumours with undefined margins and diffuse spread Spread underneath the retina and/or within the vitreous body (more than 3 mm in diameter), partial or complete retinal detachment	Systemic chemotherapy Intra-arterial chemotherapy Intravitreal chemotherapy (followed by consolidation) Enucleation
	Massive tumours taking up more than 50 % of the eyeball and/or disease-related secondary glaucoma, contact with the lens, bleeding, shrinking of eyeball etc.	(Intra-arterial chemotherapy) Enucleation; in histological high-risk situations followed by adjuvant systemic chemotherapy

The ABC classification primarily serves to assess whether an eye-preserving treatment is feasible or if enucleation is required. While tumour stage E usually includes enucleation, local therapies and/or chemotherapy are feasible for retinoblastomas of stages A to C. As for tumour stage D, it depends on certain factors (such as unilateral versus bilateral retinoblastoma) whether chemotherapy and eye-preserving strategy is an option or if it needs enucleation (see *chapter „Treatment“*).

## 2. Classification of extraocular retinoblastoma (IRSS staging system)

If retinoblastoma has spread beyond the retina, tumour removal (enucleation) is followed by classification according to the International Retinoblastoma Staging System (IRSS). This considers in particular whether the tumour has infiltrated the choroid, the dermis and/or the optic nerve, or whether it has spread into the orbit, the central nervous system (meningeosis) and/or even has distant metastases. Depending on the diagnosis, four stages are differentiated: IRSS I-IV. A retinoblastoma that won't be enucleated is assigned to stage 0 (see *table below*).

**Classification according to the „International Retinoblastoma Staging System (IRSS)“**

Stadien	Definition
0	Patient receives conservative treatment, meaning not undergoing enucleation
I	Eye enucleated, histology shows complete tumour removal



<b>Stadien</b>	<b>Definition</b>
II	Eye enucleated, histology shows complete tumour removal
III a b	Regional spread: into orbit into regional lymph nodes
IV a b	Metastases: via the blood stream (without CNS-involvement) into the CNS

The IRSS classification primarily serves as a basis for decision-making regarding adjuvant therapy following enucleation.



# Bibliography

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