Dr Steven Lapere
MBChB, DA(SA), FCOphth, MMed (UCT)
Pr 0705853

Dr Hamzah Mustak
MBChB, FCOphth, MMed (UCT)
Pr 0685917

Dr Raoul Scholtz
MBChB (UCT), DCH (SA), FCS (SA)
Pr 2603721

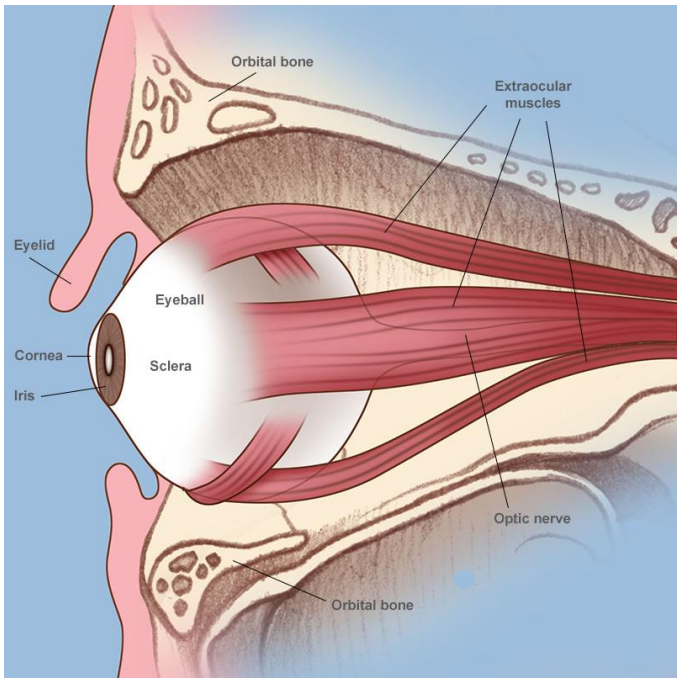
Cape Town Ocular Oncology Service: 021 671 5154 • info@ocularoncology.co.za • www.ocularoncology.co.za

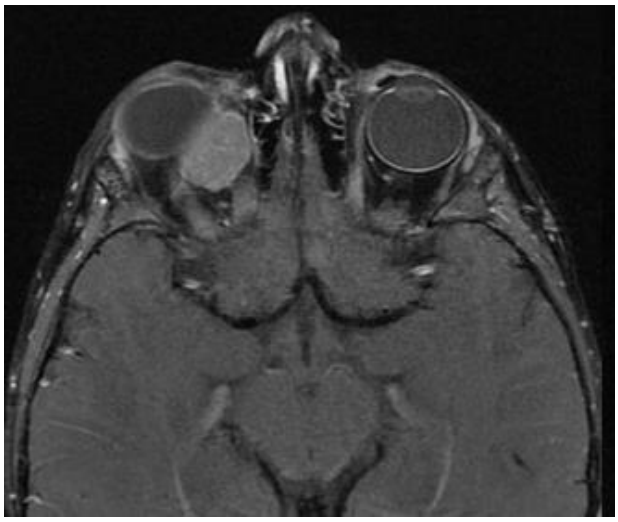
CAPE TOWN OCULAR ONCOLOGY SERVICE

Rhabdomyosarcoma

What is rhabdomyosarcoma?

Rhabdomyosarcoma is a type of sarcoma which is a cancer of the soft tissue (muscle, tendons, cartilage and bone). Rhabdomyosarcoma of the orbit usually begins in the muscles that help the eyeball move. Overall rhabdomyosarcoma is an uncommon tumour accounting for 3-4% of all childhood cancers. It is however, the most common type of soft tissue sarcoma in children and can occur in any muscle of the body.





What are the different types of rhabdomyosarcoma?

There are 3 main types of rhabdomyosarcoma based on the appearance of the cells.

- Embryonal – the most common type of rhabdomyosarcoma accounting for 65% of cases and occurs most often in the head and neck region
- Alveolar – tends to occur most often in the muscles of the arms, legs and trunk. It accounts for 25% of cases

Dr Steven Lapere
MBChB, DA(SA), FCOphth, MMed (UCT)
Pr 0705853

Dr Hamzah Mustak
MBChB, FCOphth, MMed (UCT)
Pr 0685917

Dr Raoul Scholtz
MBChB (UCT), DCH (SA), FCS (SA)
Pr 2603721

Cape Town Ocular Oncology Service: 021 671 5154 • info@ocularoncology.co.za • www.ocularoncology.co.za

CAPE TOWN OCULAR ONCOLOGY SERVICE

- Anaplastic – the least common type of rhabdomyosarcoma. Certain classification systems divide this group into botryoid, spindle cell and undifferentiated variants

What are the signs of orbital rhabdomyosarcoma?

The orbit is a narrow, cone shaped space surrounded by bone. The orbit can not easily expand and so anything that exerts a mass effect, such as a tumour will cause compression of the nerves and vessels within the orbit. The most common features are:

- Progressive pushing forward of the eye
- Limitation of eye movements
- Gradual decrease in vision
- Pain and swelling of the eyelids
- A rapidly growing lesion on the eyelids

How is the diagnosis made?

A detailed history of symptoms and examination is first performed looking for evidence of an orbital mass. The next step is to then define the lesion with an imaging study, either a CT or MRI scan of the head and orbits.
Once the location and imaging features are identified a definitive diagnosis needs to be made which can only be done with a tissue biopsy. A tissue biopsy will also tell us what type of rhabdomyosarcoma is present.

How is the biopsy taken?

A biopsy can be either incisional (part of the tumour is taken for diagnosis) or excisional (entire tumour is removed). Incisional biopsies are mostly performed on tumours that would be difficult to remove entirely. Where possible the surgeon will try to remove the whole tumour. Recent studies, however, have shown that in orbital rhabdomyosarcoma there is no difference in survival between incisional and excisional biopsies. Incisional biopsies are increasingly being performed by surgeons to limit the risk of damaging nerves and vessels with attempted excision. The eyeball is NOT removed during the biopsy. Incisions are made to the sides of the eye (eyelid or conjunctiva) that allows for the best access to the tumour.

What happens after the biopsy shows rhabdomyosarcoma?

Once the diagnosis is made, a combined team consisting of the ophthalmologist and the oncologist will make decisions regarding further care. Further treatment

Dr Steven Lapere
MBChB, DA(SA), FCOphth, MMed (UCT)
Pr 0705853

Dr Hamzah Mustak
MBChB, FCOphth, MMed (UCT)
Pr 0685917

Dr Raoul Scholtz
MBChB (UCT), DCH (SA), FCS (SA)
Pr 2603721

Cape Town Ocular Oncology Service: 021 671 5154 • info@ocularoncology.co.za • www.ocularoncology.co.za

CAPE TOWN OCULAR ONCOLOGY SERVICE

depends on a few factors, such as presence of residual tumour, type of rhabdomyosarcoma and whether there is any metastatic spread of the disease. Several further imaging and blood tests will be performed as part of a surveillance for tumours in other parts of the body. Treatment generally consists of a combination of surgery, chemotherapy and radiation treatment.
Primary rhabdomyosarcoma of the orbit is considered a localised form of the disease. Staging is further categorised on a post-surgical grouping based on the amount of residual tumour. Treatment recommendations are guided by the Intergroup Rhabdomyosarcoma Studies (IRSG). Treatment is based on grouping and is summarised in the table below.

Group	Description	Treatment
I	No residual tumour, complete excision with clear margins	Chemotherapy only
II	Microscopic residual tumour present	Chemotherapy + low dose radiotherapy
III	Macroscopic residual tumour present	Chemotherapy + high dose radiotherapy
IV	Metastatic involvement of other sites	Treatment is tailored by the oncologist with the aim of optimising quality of life

What is the prognosis of orbital rhabdomyosarcoma?

Improvements in treatment with chemotherapy regimens and radiation has greatly improved outcomes compared to the aggressive surgical approach used prior to this. The IRSG reported 3-year failure-free (no tumour recurrence) survival rates are 91% (group I), 94%(group II), 80%(group III)