Welcome to this NDRS training module on Tumours of the Eye & surrounding structures. This module is designed to help Cancer Administration staff gain a better understanding of these tumours and the terminology used by the clinical teams.

Tumours of the Eye & Adnexal Structures

- For recording purposes, tumours of the eye & adnexal structures are grouped under Brain & Central Nervous System
- For clinical purposes, tumours of the eye & adnexal structures may be covered by a different MDT depending on tumour location, morphology and stage. This may include:
  - Head & Neck MDT
  - Sarcoma MDT
  - Skin MDT
  - Neuroendocrine MDT
  - CTYA or relevant Paediatric MDT
  - Brain & CNS MDT

While tumours of the eye and surrounding structures may be covered by any of the MDTs shown - depending on the location, morphology and stage - it should be noted that they are always recorded as a subset of the Brain and Central Nervous System.

Agenda

- Eye and Adnexal Structures
  - Introduction
  - Anatomy & Physiology
  - Diagnosis
  - Treatment
- Summary
- Acknowledgements

This module may be paused at any time

In this module we'll give you a brief introduction to tumours of the eye and the areas around the eye, including some of the symptoms that patients might experience. We'll look at the anatomy & physiology of the eyes and will then go through the diagnosis and treatment options for these tumours. Remember, this module can be paused at any time.

Eye - Introduction

In this section we will cover:

- Causes and risk factors
- Signs and symptoms

We’re going to start by looking at causes and risk factors

Causes & Risk Factors – Eye

The risk factors for many eye cancers vary depending on the morphology of the tumour but broadly speaking the following may increase the risk of some eye tumours:

- Ultra-violet radiation exposure
- Having light-coloured eyes or fair skin
- Age, depending on tumour type
- Inherited cancer syndromes (BAP1 syndrome can increase the risk for melanomas, RB1 can increase the risk of retinoblastoma)
- Compromised or suppressed immune system
- Some auto-immune diseases such as rheumatoid arthritis

Different morphologies of tumour are aligned with specific risk factors – some tumours are more likely if the individual has fair skin and light-coloured eyes while genetic factors can be a risk for melanoma or retinoblastoma

Signs & Symptoms – Eye

The early symptoms of an eye tumour are most likely to be detected during a routine eye test:

- Complete or partial loss of sight in the affected eye
- A pale raised lump on the surface of the eyeball or in the surrounding tissue
- Blurred vision
- Loss of peripheral vision
- A dark spot on the iris that's getting larger
- Bulging of one eye
- Other changes in the appearance of the eye such as a drooping eyelid

Many tumours of the eye will present with symptoms that can include effects on the patient's sight in one eye, a dark spot on the iris or bulging of the affected eye.

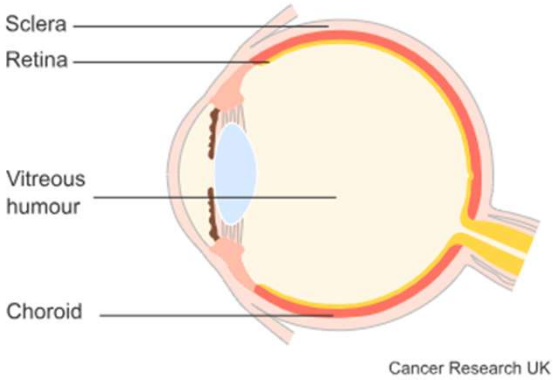
Eye - Anatomy & Physiology

In this section we will cover:

- The Eye
- Regional lymph nodes for the Eye

Now we're going to take a closer look at the eye and surrounding structures

The Eye



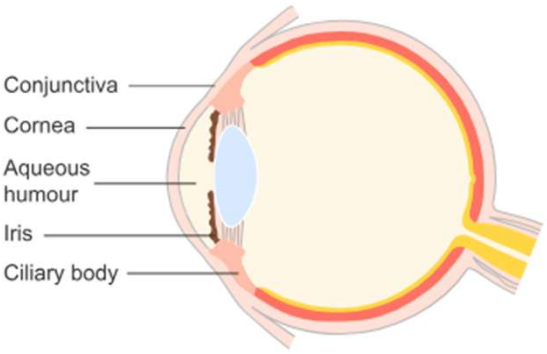
The eyeball has three layers forming the surface:

- An outer white fibrous layer (the sclera)
- A middle blood-rich layer (the choroid)
- The inner pigmented layer (the retina) – this layer reacts to light and transmits images to the brain via the optic nerve

Inside the eyeball is a clear jelly-like substance called vitreous humour which maintains the shape of the eye

The eyeball itself is formed of a three-layered outer casing. Inside the eyeball is a jelly-like substance called vitreous humour which maintains the shape of the eyeball.

The Eye



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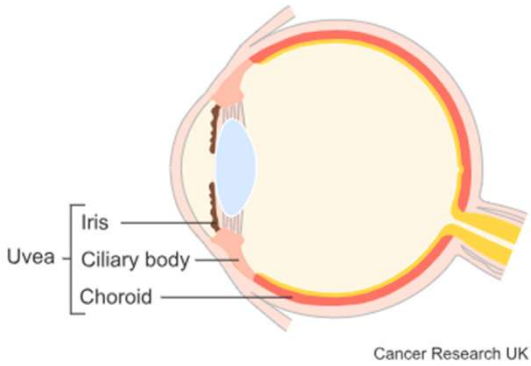
At the front of the eyeball:

- A clear, moist membrane (conjunctiva)
- Below the conjunctiva, the cornea which covers the iris and pupil
- The iris that controls how much light enters the eye by enlarging or reducing the size of the pupil
- The ciliary body is the muscle that both controls the focusing of the eye and produces the clear fluid (aqueous humour) that fills the front of the eye

Light comes into the eyeball through the pupil, which is the gap in the middle of the iris, and then through the lens which focuses the image onto the back of the eye. The size of the pupil is controlled by the ciliary body which also controls how the lens is stretched and contracted to adjust its focus.

The Eye



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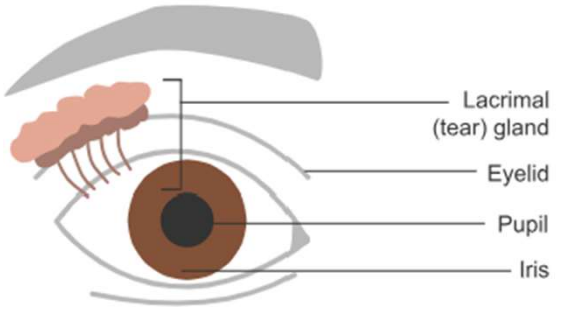
The middle of the eye (also known as the uvea) is comprised of:

- The iris
- The ciliary body
- The choroid

The uvea is the part of the eye where most cancers arise

The part of the eye known as the uvea is comprised of the iris, the ciliary body and the blood-rich middle layer of the outer casing (the choroid). The uvea is where most eye tumours arise.

The Eye



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Structures around the eye include:

- Eyelids
- Lacrimal gland that produces tears

These are known as accessory or adnexal structures to the eye

Cancers that form in these structures are often referred to as adnexal cancers

Structures surrounding the eye include the eyelid and tear ducts. These are known as accessory or adnexal structures to the eye.

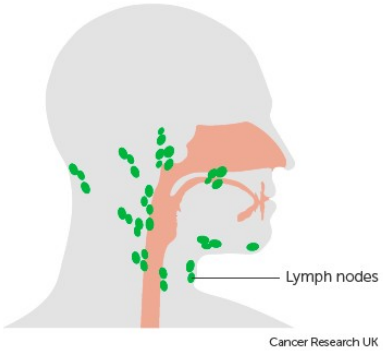
Eye – Regional Lymph Nodes

Intraocular tumours (those occurring within the eyeball) do not have any specified lymph nodes

The lymph nodes regarded as regional for other tumours of the eye and adnexal structures are ipsilateral (on the same side as the primary tumour)

Contralateral nodes (on the opposite side of the body to the primary tumour) are regarded as distant

- Pre-auricular (parotid)
- Submandibular
- Cervical



During an MDT, clinical teams will often make reference to particular groups of regional lymph nodes. This may indicate that the stage of the cancer has been determined. While tumours within the eyeball do not have specified lymph nodes, the regional lymph nodes for other tumours of the eye and adnexal structures include the pre-auricular, submandibular and cervical nodes

Eye - Diagnosis

In this section we will cover:

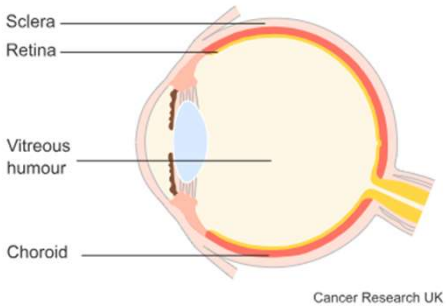
- Investigations
- Morphology
- Topography
- Stage

We'll now look at the diagnostic process

Investigations – Eye

Eye

- Eye examination
- Ultrasound scan
- Fluorescein angiogram (FA)
- Biopsy of the eye
- Blood tests
- MRI, CT or other imaging
- Lumbar puncture
- Bone marrow test
- Liver ultrasound
- Genetic testing



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Diagnostic test for a tumour of the eye may include physical examination, imaging and biopsies. A lumbar puncture, bone marrow test or genetic testing may be required in addition depending on earlier findings.

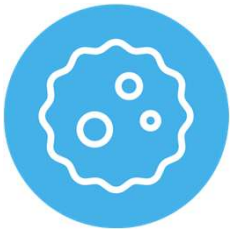
Morphology – Eye

Most malignancies of the eye are:

- Malignant melanomas – M8720/3 – other subtypes may occur, please refer to the pathology report
- Lymphoma of the eye – various morphologies

- Rarer morphologies include:
  - Retinoblastoma – M9510/3
  - Retinoblastoma, differentiated – M9511/3
  - Retinoblastoma, undifferentiated – M9512/3
  - Medulloepithelioma – M9501/3
  - Rhabdomyosarcoma – M8900/3 – (a rare childhood cancer that can occur in the orbit of the eye)

  (all of the above five morphologies are rare cancers affecting children, see the NDRS training module CTYA – Key Points https://digital.nhs.uk/ndrs/data/cancer-data-training-materials)
  - Squamous cell carcinoma – M8070/3



Most malignancies in the eye are either melanomas or lymphomas. Other, rarer morphologies include squamous cell carcinomas and a number of tumour types that primarily occur in younger patients.

Topography – Eye - Invasive ICD10

Eye

- C69.0 – Conjunctiva
- C69.1 – Cornea
- C69.2 – Retina
- C69.3 – Choroid
- C69.4 – Ciliary body
- C69.5 – Lacrimal gland and duct
- C69.6 – Orbit (not including the orbital bone – C41.0). C69.6 also applies to sarcoma of the orbit
- C69.8 – Overlapping lesion of eye and adnexa
- C69.9 – Eye, unspecified

Eye

- C49.0 – Connective and soft tissue of head, face and neck (inc. ear & eyelid) – applies to rhabdomyosarcoma and other soft tissue sarcomas of the adnexal structures – please refer to the Sarcoma training module: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Lymphoma of the eye is ICD 10 coded according to the type of lymphoma. Please refer to the Lymphoma training module: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials

With the exception of primary Lymphoma and Sarcomas, ICD10 codes for tumours of the eye are largely topographical

Topography – Eye - Non-invasive ICD10

Eye

- D03.1 – Melanoma in situ of eyelid, including canthus

Tumours of uncertain or unknown behaviour are classified with a D prefix in ICD10. Please note that melanoma-in-situ of the eyelid is registrable for COSD and would need a COSD record in your cancer data management system.

Stage – Eye

- Lymphomas of the eye are staged according to the age of the patient and the type of lymphoma. For details on staging lymphomas, please see the Lymphoma training module: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Other invasive tumours of the eye & adnexal structures are staged as follows:
    - **For diagnosis dates up to 31st December 2025 use UICC TNM v8**
    - **For diagnosis dates from 1st January 2026 use UICC TNM v9**
- Please note that the TNM version must be accurately recorded – if you are unable to amend the version on your cancer data management system, please refer to your line manager
- If, after 1st January 2026, your cancer data management system has not been amended to include TNM v9 please add the following statement to the Primary Diagnosis Subsidiary Comment field: **Patient staged using TNM9 not TNM8 as per CR2070**
- For details on recording stage, please see the NDRS training modules KPI-TNM Staging 101 and KPI-Haematology Staging 101, available on this link: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials

Lymphomas are staged according to the type of lymphoma and the age of the patient – please refer to the lymphoma training module for details. Other invasive tumours of the eye and adnexal structures are staged using the appropriate version of UICC TNM stage.

Stage – Eye

- Staging data sheets for clinical use are available from the NDRS website: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets
- Retinoblastoma require additional staging assessment, also available on the NDRS website: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets/retinblastoma
- For more details on retinoblastoma, please see the CTYA Key Points training module: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Please be aware that different morphologies may require a different staging data sheet

Staging parameters and details on recording different types of tumour can be found on the NDRS website

Eye - Treatment

In this section we will cover:

- Surgery
- Radiotherapy
- Chemotherapy
- Palliative

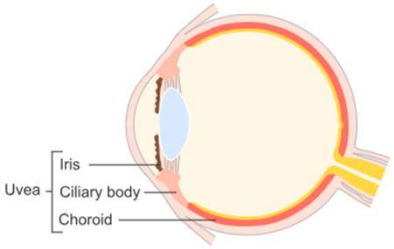
Treatment for tumours in and around the eye will vary depending on the type, stage and location of the tumour.

Monitoring – Eye

Monitoring

Some tumours of the eye, such as a melanomas of the iris, choroid or ciliary body may be very slow growing and may not cause any symptoms

Clinicians may recommend a conservative approach consisting of regular monitoring to ensure the tumour is not growing or causing issues



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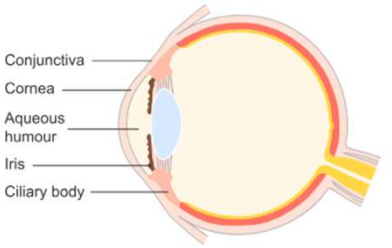
A conservative approach may be recommended for some areas of the eye where the tumour is asymptomatic and slow growing

Surgery – Eye

Surgery

- A melanoma in the iris may be small enough to resect without removing the eyeball
- If a tumour in the eyeball is large or seriously affecting vision, the eyeball may be surgically removed – this surgery is known as an enucleation
- If there is cancerous spread beyond the eyeball, surrounding tissue may also be removed in a surgery known as an orbital exenteration
- If a recurrent melanoma in the eyeball is diagnosed, enucleation may be offered even if the recurrence is small



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… but if surgery is necessary, very small tumours in the iris may be resectable without removing the eye. Larger, symptomatic or recurrent tumours may require the removal of the eyeball, possibly including the excision of surrounding tissue depending on the spread of the tumour.

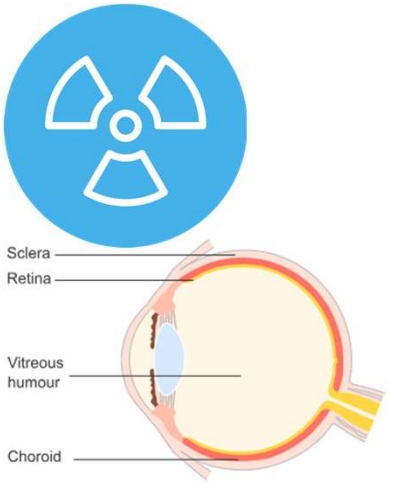
Radiotherapy – Eye

Radiotherapy

- Radiotherapy may be offered as a primary treatment or as an adjuvant treatment depending on the type, location and stage of the tumour

Radiotherapy – External beam

- The methods of administering external beam radiotherapy include:
  - Proton therapy – while most forms of radiotherapy use x-ray radiation, proton therapy uses positively charged particles delivered at high speed to destroy cancer cells
  - Stereotactic radiosurgery – this method uses a large number of very narrow radiation beams to deliver high dose radiotherapy into the tumour

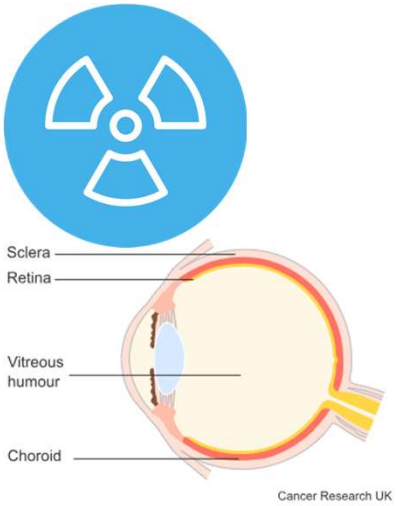


Radiotherapy may be offered as a primary treatment or adjuvantly, depending on the type, stage and location of the tumour. Radiotherapy may be delivered externally, either as proton therapy or as stereotactic radiosurgery…

Radiotherapy – Eye

Radiotherapy - Brachytherapy

- The method of administering brachytherapy to eye tumours is known as plaque radiotherapy
  - A small radioactive disc is surgically attached to the surface of the eyeball, directly over the tumour
  - The plaque is left in place for a few days then surgically removed



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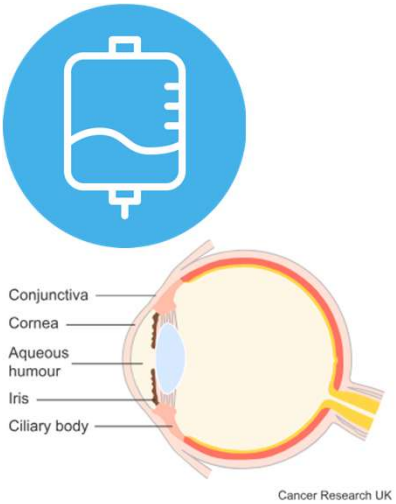
… or it may be delivered as brachytherapy utilising a surgically attached radioactive disc on the surface of the eye.

Chemotherapy – Eye

Chemotherapy

- Chemotherapy is unlikely to work for uveal melanoma as a primary treatment but is sometimes offered as an adjuvant treatment
- Chemotherapy in the form of eye drops may be offered for conjunctival melanoma or conjunctival SCC
- Chemotherapy is most frequently offered for lymphoma of the eye



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Chemotherapy is usually offered for lymphoma of the eye but may be offered for other tumour morphologies

Summary

In Summary…

Summary

- Tumours of the eye and adnexal structures are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used

The diagnosis of tumours of the eye and surrounding structures normally relies on imaging but may include pathological examination and molecular testing.

Summary

- Tumours of the eye and adnexal structures are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Primary lymphomas of the eye and adnexal structures require the relevant site-specific stage to be recorded. Other invasive tumours are staged using the appropriate UICC TNM version

Primary lymphomas must be staged using the relevant site-specific staging system. Other invasive tumours are staged using the appropriate UICC TNM version

Summary

- Tumours of the eye and adnexal structures are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Primary lymphomas of the eye and adnexal structures require the relevant site-specific stage to be recorded. Other invasive tumours are staged using the appropriate UICC TNM version
- Surgery may be offered depending on tumour type, stage and location. Resection may be possible but an advanced tumour in the eye may require the removal of the eyeball (enucleation)

Surgery may be offered where appropriate depending on tumour type, stage and location

Summary

- Tumours of the eye and adnexal structures are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Primary lymphomas of the eye and adnexal structures require the relevant site-specific stage to be recorded. Other invasive tumours are staged using the appropriate UICC TNM version
- Surgery may be offered depending on tumour type, stage and location. Resection may be possible but an advanced tumour in the eye may require the removal of the eyeball (enucleation)
- Radiotherapy may be offered, either as external beam radiation or as a form of brachytherapy known as plaque radiation

If radiotherapy is offered, it may be as external beam radiation or as a particular type of brachytherapy

Summary

- Tumours of the eye and adnexal structures are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Primary lymphomas of the eye and adnexal structures require the relevant site-specific stage to be recorded. Other invasive tumours are staged using the appropriate UICC TNM version
- Surgery may be offered depending on tumour type, stage and location. Resection may be possible but an advanced tumour in the eye may require the removal of the eyeball (enucleation)
- Radiotherapy may be offered, either as external beam radiation or as a form of brachytherapy known as plaque radiation
- Chemotherapy is most often used for lymphomas but may be offered for other morphologies depending on type, stage and location

While chemotherapy is usually offered for lymphomas, it may also be offered for other morphologies.

Summary

- If in any doubt as to whether you should be recording a diagnosis, please refer to the latest COSD User Guide, Appendices A, B & C
- For guidance on the required staging system, please refer to the latest COSD User Guide, Appendix E

- https://digital.nhs.uk/ndrs/data/data-sets/cosd#downloads

Do please remember, guidance **is** available on our website. You can download the COSD User Guide by clicking on this link and selecting the COSD version appropriate to your trust.

Acknowledgements

Many thanks to Cancer Research UK for the use of their images within this training module.

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