Welcome to this NDRS training module on Tumours of the Brain and Central Nervous System. This module is designed to help Cancer Administration staff gain a better understanding of these tumours and the terminology used by the clinical teams.

Agenda

- Brain and Spinal Cord
  - Introduction
  - Anatomy & Physiology
  - Diagnosis
  - Treatment
- Summary
- Acknowledgements

This module may be paused at any time

We're going to give you a brief introduction to these tumours including some of the symptoms that patients might experience. We'll look at the anatomy & physiology of the brain & spinal cord and then go through the diagnosis and treatment options. Remember, this module can be paused at any time.

Brain & CNS - Introduction

In this section we will cover:

- Causes and risk factors
- Signs and symptoms

We’re going to look first at the causes and risk factors

Causes & Risk Factors – Brain & CNS

Causes of brain tumours are mostly unknown but there are a few known risk factors:

- Age – more common in children and older adults, depending on the tumour type
- Obesity – increased risk of meningioma
- Radiation – previous exposure to ionising radiation
- Family History and Genetics – some genetic syndromes increase the risk of brain tumours

The cause of many Brain & CNS tumours is largely unknown but some *known* risk factors include: age, being very overweight, and prior exposure to medical radiation. Certain genetic syndromes have *also* been linked with brain and CNS tumours.

Signs & Symptoms - Brain

Intracranial pressure

- Headaches in the morning
- Seizures / Fits
- Nausea and vomiting
- Personality changes
- Memory defect
- Intellectual impairment
- Drowsiness / lethargy

In the case of brain tumours, intracranial pressure alone may cause a number of symptoms. The skull is a finite space and additional tissue within that space can often put pressure on other brain structures. Symptoms can include headaches, seizures, personality changes and memory problems.

Signs & Symptoms - Brain

Intracranial pressure

- Headaches in the morning
- Seizures / Fits
- Nausea and vomiting
- Personality changes
- Memory defect
- Intellectual impairment
- Drowsiness / lethargy

Tumour Location

- Unilateral Weakness
- Visual disturbances
- Language problems
- Co-ordination problems
- Vertigo
- Difficulty swallowing
- Problems with smelling and hearing

Presenting symptoms may give clues about the location of the tumour as different parts of the brain deal with different processes.

Signs & Symptoms – Spinal Cord

- Back and neck pain
- Numbness
- Tingling and weakness in either the arms or legs
- Cauda equina syndrome (dysfunction of multiple lumbar and sacral nerve roots at the base of the spine)
- Clumsiness
- Difficulty in walking
- Incontinence - loss of control of the bladder and bowel

In the Spinal cord, signs of a possible tumour may include numbness, tingling sensations in the limbs, difficulty walking or a loss of bladder or bowel control

Brain & CNS - Anatomy & Physiology

In this section we will cover:

- The Nervous System
- The Brain
- The Spinal Cord

It’s helpful to have an understanding of how the Brain and spinal cord function.

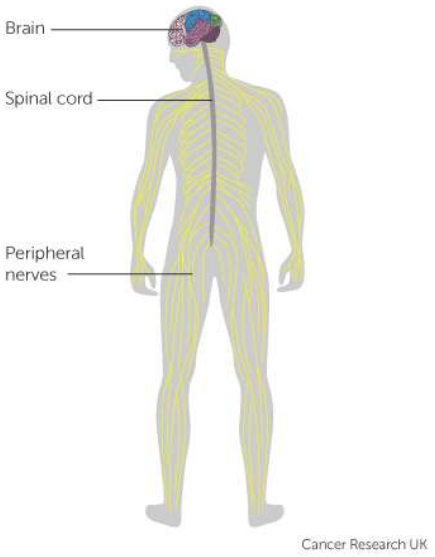
The Nervous System

- The nervous system is the major controlling, regulatory, and communicating system in the body
- Together with the endocrine system, the nervous system is responsible for regulating and maintaining homeostasis (a state of balance within the body's systems)
- Through its receptors, the nervous system keeps us in touch with our environment, both external and internal

The nervous system receives and sends information that helps it to keep our bodily systems in a state of balance

The Nervous System

- **Central Nervous System** (CNS) consists of the brain and spinal cord
- **Peripheral Nervous System** (PNS) consists of all the sensory nerves that feed information to the spinal cord and brain from the rest of the body and motor nerves which carry messages to other parts of the body from the brain and spinal cord



It's classified as either the Central Nervous System – which is comprised of the brain and spinal cord – or the Peripheral Nervous System – which describes the nerves in the rest of the body.

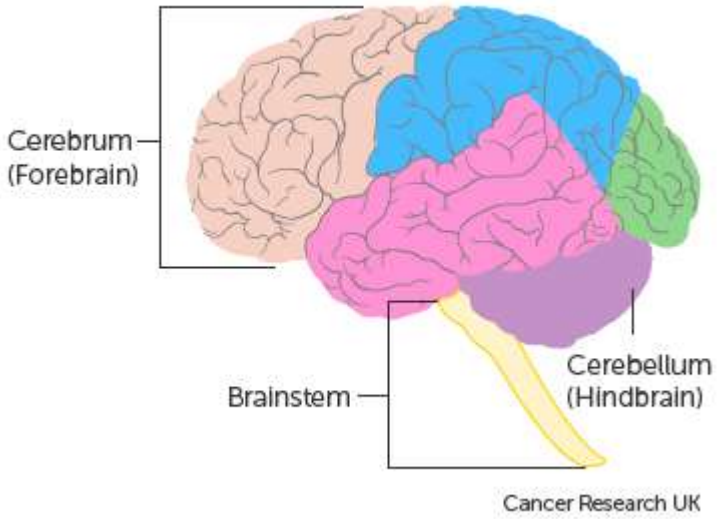
The Brain

The brain is comprised of :

The Cerebrum (Forebrain)

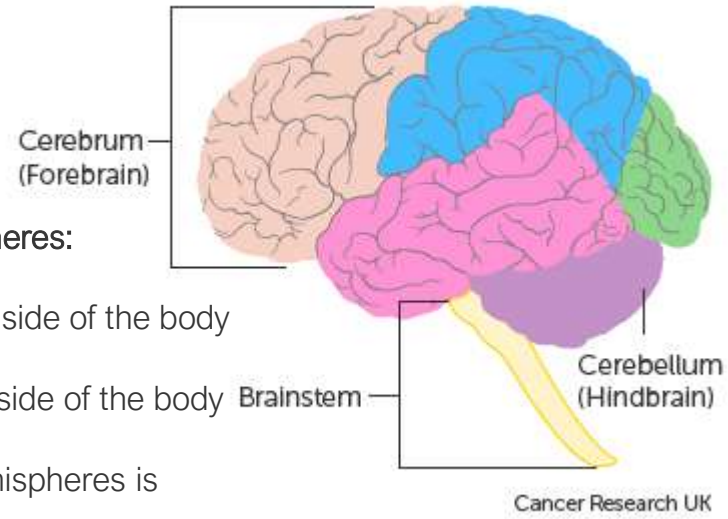
The Cerebellum (Hindbrain)

The Brainstem



Broadly speaking, the brain is composed of three major parts: The cerebrum or forebrain, the cerebellum, also known as the hindbrain, and the brainstem.

The Brain



The Cerebrum is split into two hemispheres:

The Left Hemisphere controls the right side of the body

The Right hemisphere controls the left side of the body

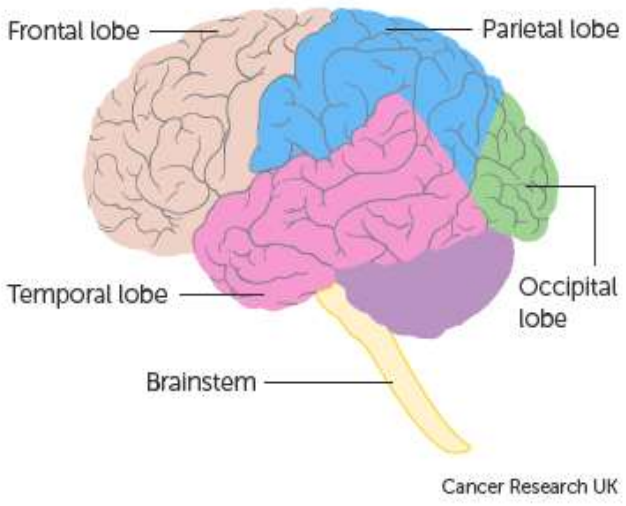
The area of the brain that joins the hemispheres is called the Corpus Callosum

The Cerebrum is split into two halves, left and right. Each half controls the opposite side of the body. The central portion of the brain that connects these halves is called the Corpus Callosum.

The Brain

Each hemisphere of the Cerebrum contains four lobes:

Frontal lobe
- Speech
- Problem solving
- Personality
- Emotional processing
- Movement initiation

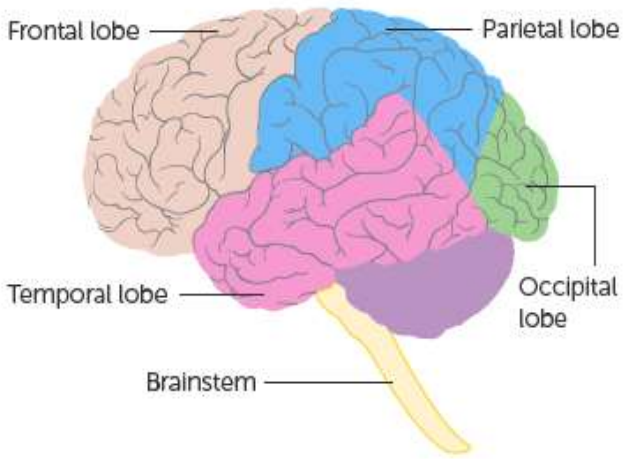


Each half of the Cerebrum is made up of four lobes. The Frontal lobe is where we deal with emotions and it's also connected to our personalities and our problem solving abilities

The Brain

Each hemisphere of the Cerebrum contains four lobes:

Temporal lobe
- Sounds
- Memory storage
- Language processing

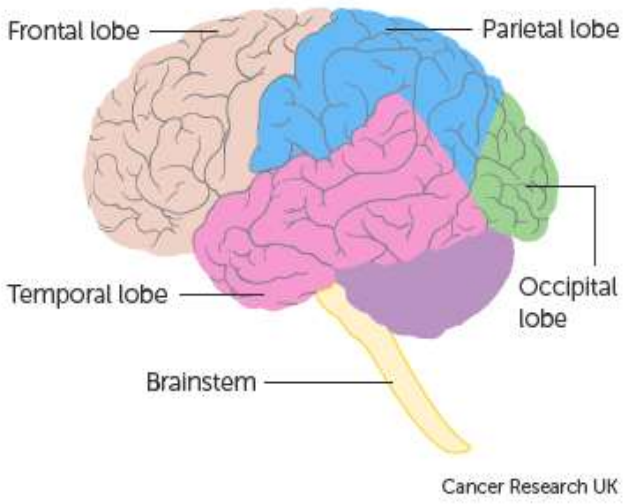


The Temporal lobe is where memories are stored and where we make sense of language

The Brain

Each hemisphere of the Cerebrum contains four lobes:

Parietal lobe
- Touch
- Temperature
- Pain
- Object recognition
- Sensory perception & integration

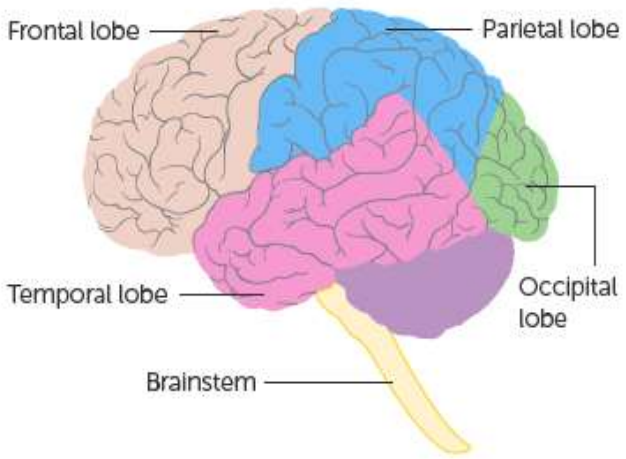


While the role of the Parietal lobe includes the processing of touch, temperature and pain …

The Brain

Each hemisphere of the Cerebrum contains four lobes:

Occipital lobe
Visual processing

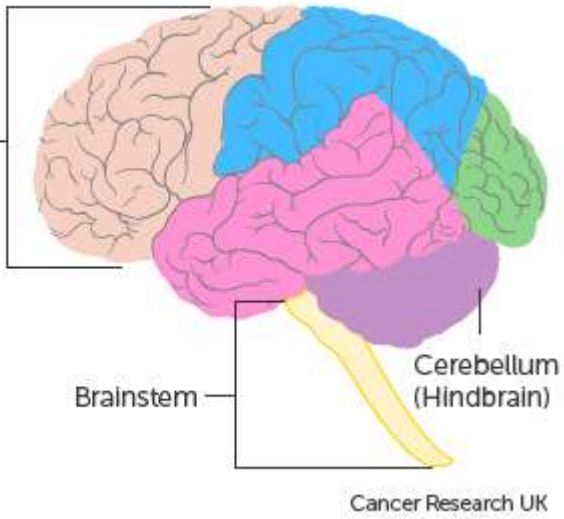


… and the Occipital lobe processes visual input

The Brain

At the back of the brain is the Cerebellum

Balance
Posture
Co-ordination



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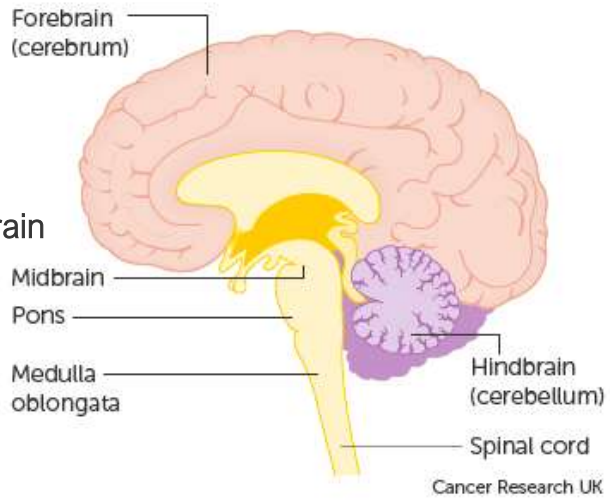
Behind the Cerebrum, the Cerebellum, or hindbrain, controls things like our sense of balance and our co-ordination…

The Brain

The Brain Stem is located at the base of the brain

- Breathing
- Heartbeat
- Blood pressure
- Coughing
- Swallowing



…while the Brain Stem controls the autonomic functions – the functions that usually happen without any conscious input – for instance, breathing and keeping the heart beating. It also controls functions such as coughing and swallowing

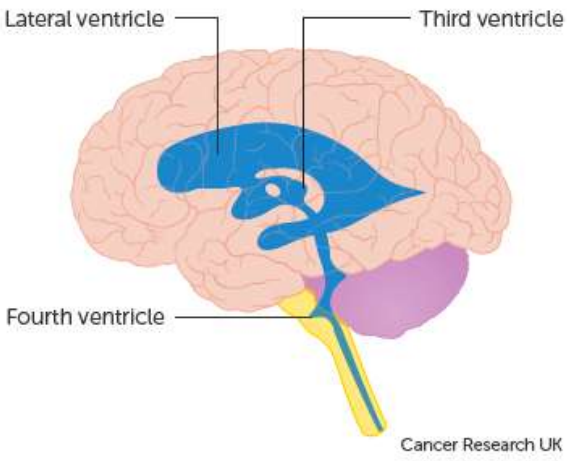
The Brain

Ventricles

Four interconnected chambers in the brain, filled with cerebrospinal fluid (CSF)

- Lateral ventricles (two) - located in each hemisphere of the cerebrum
- Third ventricle - a narrow cavity superior to the hypothalamus and between the right and left halves of the thalamus
- Fourth ventricle - lies between the brain stem and the cerebellum

The Choroid plexus is a separate structure within the ventricles which produces the cerebrospinal fluid



In the central portion of the brain, above the brainstem, lie the ventricles which are filled with cerebrospinal fluid, sometimes called CSF. CSF is produced by the Choroid plexus, a separate structure within the ventricles.

The Brain

- Deep within the brain are the Pituitary gland and the Pineal gland
  - The Pituitary gland affects growth, metabolic rate, egg / sperm production
  - The Pineal gland produces melatonin which regulates sleep

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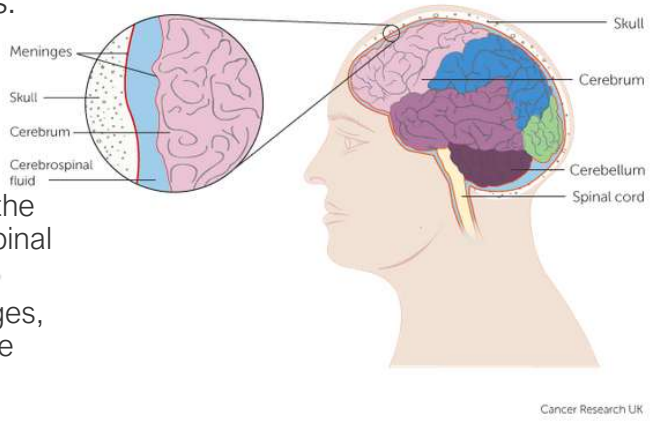
In front of the ventricles, the Pituitary gland controls growth, metabolism and the production of egg cells or sperm cells. Located just behind the ventricles, the Pineal gland is responsible for regulating our sleep cycle.

The Brain

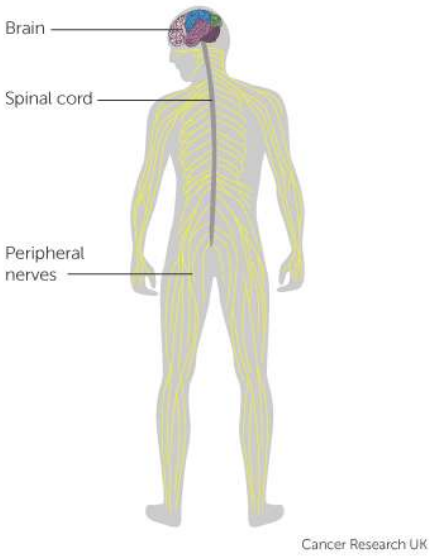
- Meninges
- A membrane covering the brain and spinal cord and it is composed of three layers:
  - Dura Mater - outermost layer
  - Arachnoid Mater - the middle layer
  - Pia Mater - innermost layer
  - Between the Arachnoid and Pia Mater, the subarachnoid space contains cerebrospinal fluid (CSF)
  - Tentorium - A specific fold of the meninges, which separates the cerebellum from the cerebrum



The brain is covered by a multi-layered membrane called the meninges. The Dura Mater is the outermost layer with the Arachnoid Mater and the Pia Mater underneath. CSF fills the space between the Arachnoid Mater and the Pia Mater and one of the functions of the CSF is to act as a cushion for the brain within the sub-arachnoid space.

The Spinal Cord

- The Spinal Cord is continuous with the brain stem and extends from the foramen magnum at the base of the skull to the level of the first lumbar vertebra
- It is divided into 31 segments with each segment giving rise to a pair of spinal nerves



The brain stem extends down the spine, where it's referred to as the spinal cord. The spinal cord gives rise to multiple pairs of spinal nerves which in turn branch out to form the rest of the peripheral nervous system.

Brain & CNS - Diagnosis

In this section we will cover:

- Investigations
- Morphology
- Topography
- Stage
- Grade

We'll now look at the diagnostic process

Investigations – Brain & CNS

- X-ray
- CT scan
- MRI scan
- PET/CT scan

- Biopsy
- Cytology – Cerebrospinal Fluid (CSF) via Lumbar puncture
- Molecular testing

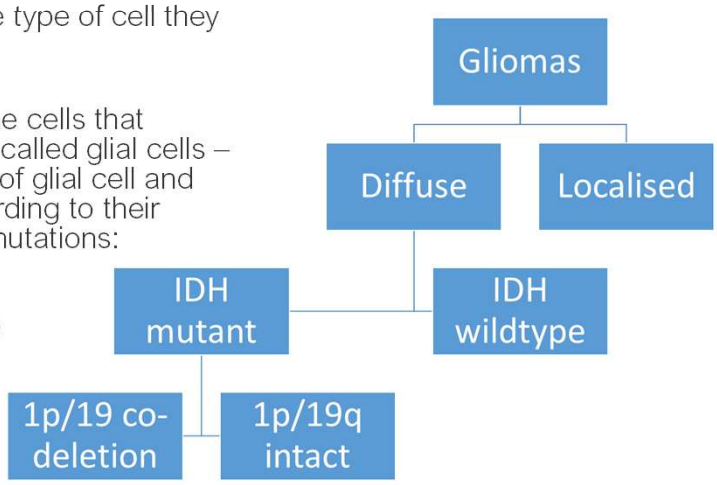
Investigations for suspected Brain or Spinal Cord tumours will usually include radiological examinations: such as X-rays or scans. Diagnosis of a tumour may require solid tissue samples from a biopsy or fluid samples for cytology. Molecular testing may also be used, both as part of the diagnostic process and to determine the most appropriate treatment.

Morphology

There are nearly 100 different types of brain tumours which are generally named after the type of cell they develop in

Most brain tumours develop from the cells that support the nerve cells of the brain called glial cells – glioma. There are many sub-types of glial cell and the tumours are often defined according to their specific cell characteristics or cell mutations:

- Astrocytic tumours (Astrocytoma)
- Oligodendrogliomas
- Mixed gliomas
- Ependymomas



There are almost 100 different types of brain tumours, too many to list here. Many of these tumours develop from glial cells – the cells that support the nerve cells within the brain. Glial cells are further divided into sub-types and as tumours are often named after the cells in which they've arisen, gliomas may have more specific names such as Oligodendroglioma or Astrocytoma. In some cases, the grade of the tumour may also affect the name: for instance, a grade 4 astrocytoma is usually referred to as a glioblastoma multiforme.

Morphology - Brain & CNS

Other cells within the brain and CNS give rise to different types of tumours. These include:

- Meningiomas – in the cells of the Meninges
- Neurilemmoma (Schwannoma) – Schwann cells: fatty cells on the outside of nerves
- Ganglioneuroma - Ganglion cells
- Embryonal tumours – in cells that are left over from our developmental stage in utero (these can include medulloblastomas and atypical teratoid/rhabdoid tumours)
- Primary cerebral (or central nervous system) Lymphoma – started with Lymphocytes within the brain (or spinal cord)
- Choroid plexus tumours – in the area of the brain ventricles where cerebrospinal fluid is formed

Due to the range of possible types / subtypes of tumour it's not possible to list all morphology codes here. Please refer to the pathology report and your clinical team as well as the SNOMED CT Browser: https://termbrowser.nhs.uk/

Tumours arising in other cell types include meningiomas, embryonal tumours (such as medulloblastomas) and primary cerebral or CNS lymphoma. When recording morphology, always refer to the pathology report.

Topography – Brain - Invasive ICD10

Meninges

- C70.0 - Cerebral Meninges
- C70.1 - Spinal Meninges
- C70.9 - Meninges NOS

Endocrine & related structures

- C75.1 - Pituitary gland
- C75.2 - Craniopharyngeal duct
- C75.3 - Pineal gland

Primary cerebral or CNS Lymphoma

- Coded to the relevant Lymphoma ICD10 code

Brain

- C71.0 - Cerebrum
- C71.1 - Frontal lobe
- C71.2 - Temporal lobe
- C71.3 - Parietal lobe
- C71.4 - Occipital lobe
- C71.5 - Ventricle, NOS (excludes fourth ventricle)
- C71.6 - Cerebellum, NOS
- C71.7 - Brain stem / fourth ventricle / Infratentorial NOS
- C71.8 - Overlapping lesion of brain
- C71.9 - Brain, NOS

With the exception of primary brain or central nervous system Lymphoma, ICD10 disease codes for Brain & CNS tumours are topographical - they describe where the tumour is. An ICD10 code prefix of C also indicates that the tumour is invasive. When assigning the ICD10 code for registrable Invasive tumours of the Brain, meninges or cranial endocrine & related structures, these are the relevant codes. Primary lymphoma of the brain or central nervous system would be ICD10 coded to the relevant lymphoma.

Topography – Benign ICD10

Meninges

- D32.0 - Cerebral Meninges
- D32.1 - Spinal Meninges
- D32.9 - Meninges NOS

Endocrine & related structures

- D35.2 - Pituitary gland
- D35.3 - Craniopharyngeal duct
- D35.4 - Pineal gland

Brain

- D33.0 – Brain, supratentorial
  - Cerebral ventricle
  - Cerebrum
  - All lobes
- D33.1 – Brain, infratentorial
  - Brain stem
  - Cerebellum
  - Fourth ventricle
- D33.2 – Brain, unspecified

… whereas D prefix codes for Benign tumours of the same areas are shown here…

Topography – Central Nervous System

Spinal Cord, Cranial Nerves, and other parts of the Central Nervous System - Invasive

- C72.0 - Spinal cord
- C72.1 - Cauda equina
- C72.2 - Olfactory nerve
- C72.3 - Optic nerve
- C72.4 - Acoustic nerve
- C72.5 - Cranial nerve, NOS
- C72.8 - Overlapping lesion of brain and central nervous system
- C72.9 - Nervous system, NOS

Primary cerebral or CNS Lymphoma

- Coded to the relevant Lymphoma ICD10 code

Spinal Cord, Cranial Nerves, and other parts of the Central Nervous System - Benign

- D33.3 - Cranial nerves
- D33.4 - Spinal cord
- D33.7 – Other specified parts of Central Nervous System
- D33.9 – Central Nervous System, unspecified

… and the codes for both Invasive and Benign spinal cord and CNS tumours are listed here.

Topography – Brain & CNS - Non-invasive ICD10

Meninges

- D42.0 - Cerebral Meninges
- D42.1 - Spinal Meninges
- D42.9 - Meninges NOS

Endocrine & related structures

- D44.3 - Pituitary gland
- D44.4 - Craniopharyngeal duct
- D44.5 - Pineal gland

Brain & Central Nervous System

- D43.0 – Brain, supratentorial
- D43.1 – Brain, infratentorial
- D43.2 – Brain, unspecified
- D43.3 – Cranial nerves
- D43.4 – Spinal cord
- D43.7 – Other parts of Central Nervous System
- D43.9 – Central Nervous System, unspecified

Tumours may also be classified as Uncertain or Unknown behaviour or, depending on the location and tumour behaviour, in-situ. These are also D prefixed in ICD10. Please note that all Brain & CNS ICD10 codes shown in this module are registrable for COSD and would need a COSD record in your cancer management system.

Stage – Brain & CNS

Most tumours of the brain and central nervous system are not considered to be stageable in TNM. The tumours that are stageable with an M0-M4 Chang stage are:

- Central nervous system germ cell tumours (these include germinomas and non-germinomas. Non-germinomas of the CNS include the sub-categories: embryonal carcinomas, endodermal sinus/yolk sac tumours and specific teratomas)
- CNS embryonal tumours, NOS
- Atypical teratoid/rhabdoid tumours (ATRT)
- Pineoblastoma
- Ependymoma

Most tumours of the brain and CNS are not considered to be stageable in TNM v8. The tumours listed here are stageable with the Chang staging system.

Stage – Brain & CNS

- Medulloblastoma
  - for diagnosis dates up to 31st December 2025 a Chang stage is required
  - for diagnosis dates from 1st January 2026, either a TNM v9 stage or a Chang stage may be submitted
    - 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 record the TNM v9 stage and add the following statement to the Primary Diagnosis Subsidiary Comment field:
    - Patient staged using TNM9 not TNM8 as per CR2070

For diagnosis dates from 1st January 2026 Medulloblastomas may be staged with either a Chang stage or UICC TNM v9.

Stage – Brain & CNS

- An M0-M4 Chang stage should be recorded for all Chang-stageable tumours. A staging data sheet for Brain & CNS - Chang Stage may be downloaded from: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets

CHANG·STAGING·SYSTEM¶

¶

STAGE¤	Description¤
M0¤	No·evidence·of·gross·subarachnoid·or·haematogenous·metastasis.¤
M1¤	Microscopic·tumour·cells·found·in·cerebrospinal·fluid.¤
M2¤	Gross·nodular·seedings·demonstrated·in·the·cerebellar,·cerebral·subarachnoid·space,·or·in·the·third·or·lateral·ventricles¤
M3¤	Gross·nodular·seeding·in·spinal·subarachnoid·space¤
M4¤	Extraneuroaxial·metastasis.¤

A Brain & CNS Chang staging sheet (for clinical use) may be downloaded from the link shown.

Grade – Brain & CNS

World Health Organization (WHO) grading system.
Captures not just differentiation and biological behaviour, but also prognostic factors, acting as guidance for treatment plans.

Grades I & II are LOW GRADE

Grade I

- Slow growing
- Less likely to recur

Grade II

- Slow growing
- Can be infiltrative & may recur.

Grade is assigned to a tumour based on the World Health Organisation grading system. Grades I and II are LOW grade…

Grade – Brain & CNS

World Health Organization (WHO) grading system.
Captures not just differentiation and biological behaviour, but also prognostic factors, acting as guidance for treatment plans.

Grades I & II are LOW GRADE

Grade I

- Slow growing
- Less likely to recur

Grade II

- Slow growing
- Can be infiltrative & may recur

Grades III & IV are HIGH GRADE

Grade III

- Malignant
- Faster growing

Grade IV

- Malignant
- Very rapid growth

… while grades III and IV are HIGH grade.

Brain & CNS - Treatment

In this section we will cover:

- Surgery
- Radiotherapy
- Chemotherapy
- Palliative

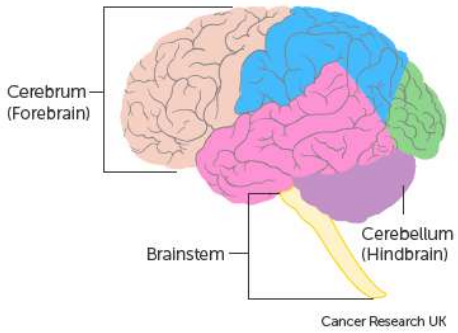
Treatment for Brain and CNS tumours varies depending on the type, size and location of the tumour.

Surgery – Brain & CNS

Surgery

Surgery can remove brain & CNS tumours, but it's very difficult to remove all tumour cells due to the delicate nature of tissues. Surgical treatments tend to be conservative to prevent serious side effects

If it's not possible to totally remove a brain tumour surgery may relieve symptoms to ensure a better quality of life for the patient

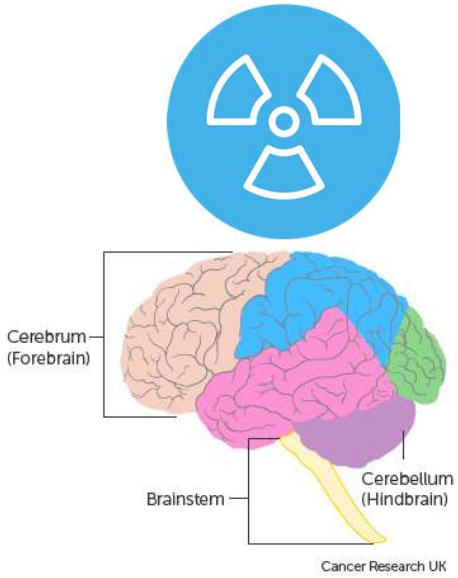


While surgery can remove tumours of the brain & central nervous system, it's difficult to remove all tumour cells without destroying surrounding tissue. However, removal of a brain tumour may relieve symptoms related to pressure inside the skull.

Radiotherapy – Brain & CNS

Radiotherapy

- Radiotherapy improves survival for high grade brain & CNS tumours. It can prove curative for tumours such as medulloblastoma and germinoma, but not in others such as glioma
- Radiosurgery gives a very high dose of radiation to a very small precise area
- Proton beam therapy is very good at treating some skull base and children's brain tumours

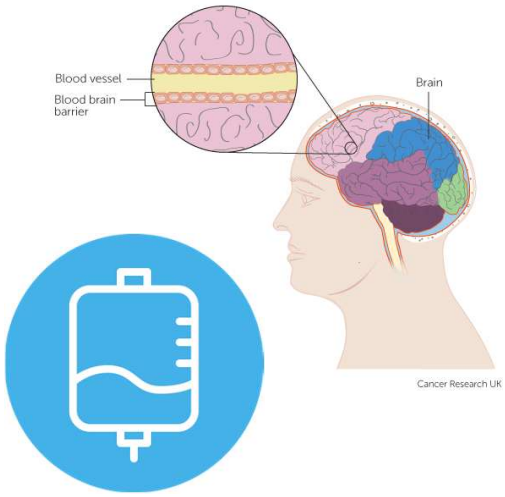


Radiotherapy can improve survival and even prove curative for some types of Brain & CNS tumour. Radiosurgery uses high dose radiation in a tightly controlled beam to minimise damage to the surrounding healthy tissue.

Chemotherapy – Brain

Chemotherapy

- It can be difficult to treat brain tumours with chemotherapy because the brain is protected by the blood-brain barrier which only allows certain substances through from the blood to the brain tissues. Only a few chemotherapy drugs can get across the blood-brain barrier and chemotherapy is most commonly given for high grade tumours



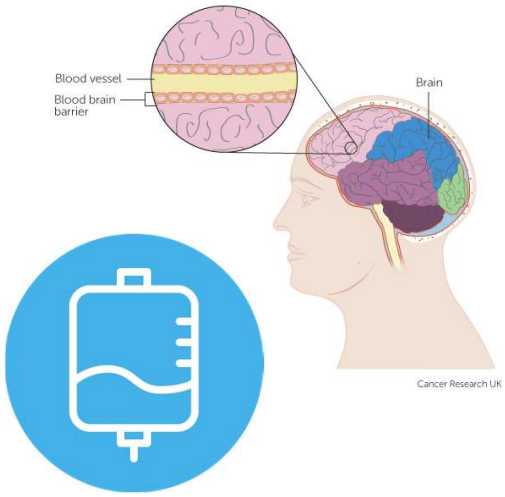
For the treatment of brain tumours, the choice of available Chemotherapy is limited. This is because many types of chemotherapy are unable to cross the blood-brain barrier within the blood vessels.

Chemotherapy

Chemotherapy

- Adjuvant chemotherapy given at the time of the primary treatment may improve survival in patients with gliomas; however, response rates are usually low

- Chemotherapy drugs in gel wafers can be place inside the brain following debulking surgery. The gel dissolves over time slowly releasing the chemotherapy drug directly into the brain



One way of delivering chemotherapy is to insert a gel wafer containing the drug directly into the brain after debulking surgery.

Palliative

Palliative

For some patients with advanced tumours no treatment is offered other than symptomatic care

Many symptoms associated with brain tumours are associated with intracranial pressure:

- A shunt may be inserted in order to relieve intracranial pressure
- Steroids are commonly prescribed to control symptoms but are not classified as 'treatment' to the tumour

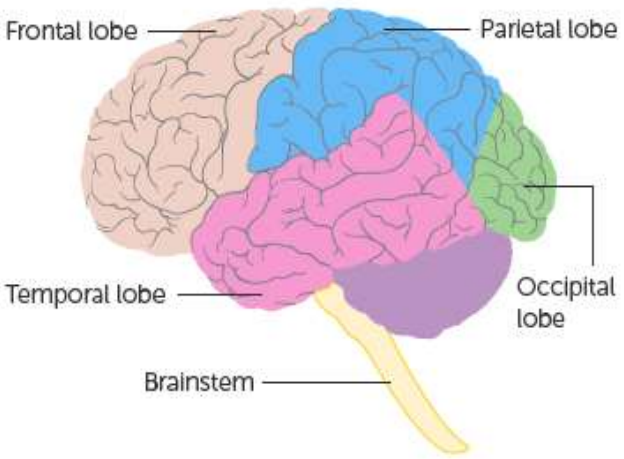
Some tumours are so advanced that only Palliative treatment is offered to relieve symptoms. This may include a shunt to relieve intracranial pressure or steroids.

In Summary

To summarise…

Summary

- The cause of many brain / CNS tumours is not known but age group, obesity, medical radiation and genetics are a factor in some

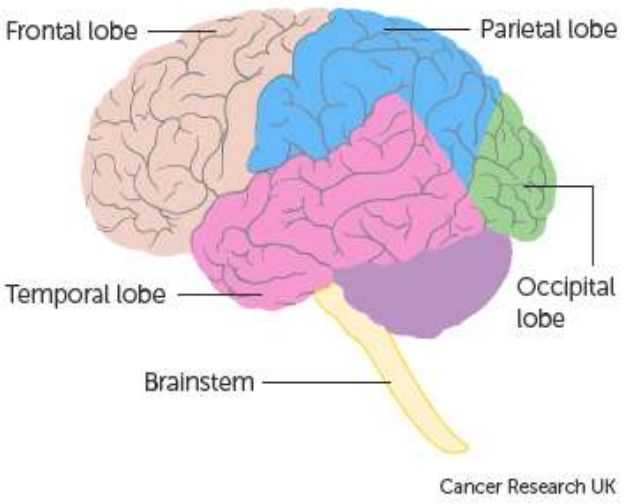


Age, obesity, medical radiation or genetics are thought to be contributory factors in some brain and CNS tumours.

Summary

- The cause of many brain / CNS tumours is not known but age group, obesity, medical radiation and genetics are a factor in some

- Different parts of the brain control different bodily functions which may be specifically affected by the presence of a tumour. The presence of a tumour will usually cause intracranial pressure to increase, leading to a worsening of symptoms



Even if benign, the presence of a tumour may cause pressure within the skull.

Summary

- Brain / CNS tumours are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used

Radiology is a common diagnostic tool for Brain & CNS tumours although histology, cytology and molecular testing are also used.

Summary

- Brain / CNS tumours are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Most Brain & CNS tumours are not considered stageable, but certain specified morphologies of tumour require a Chang stage

While most brain & CNS tumours are not considered stageable, some tumour types require a Chang Stage

Summary

- Brain / CNS tumours are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Most Brain & CNS tumours are not considered stageable, but certain specified morphologies of tumour require a Chang stage
- Some brain / CNS tumours are treated with surgery. This may be to debulk the tumour, but complete removal can be difficult due to the risk of damage to surrounding tissue

Surgery may be used to debulk a tumour but there is a risk of damage to surrounding tissue.

Summary

- Brain / CNS tumours are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Most Brain & CNS tumours are not considered stageable, but certain specified morphologies of tumour require a Chang stage
- Some brain / CNS tumours are treated with surgery. This may be to debulk the tumour, but complete removal can be difficult due to the risk of damage to surrounding tissue
- Radiotherapy works well with some tumours and may be applied in the form of high dose Radiosurgery

Some tumours respond well to radiotherapy but not all.

Summary

- Brain / CNS tumours are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Most Brain & CNS tumours are not considered stageable, but certain specified morphologies of tumour require a Chang stage
- Some brain / CNS tumours are treated with surgery. This may be to debulk the tumour, but complete removal can be difficult due to the risk of damage to surrounding tissue
- Radiotherapy works well with some tumours and may be applied in the form of high dose Radiosurgery
- The range of Chemotherapy available is limited due to the effect of the blood-brain barrier. Some Chemotherapy drugs can be inserted directly into the brain in the form of a gel wafer

The blood-brain barrier limits the chemotherapy types available for use with Brain tumours. Certain types of chemotherapy gel wafers can be inserted directly into the brain where appropriate.

Summary

- Brain / CNS tumours are normally diagnosed using radiological examination +/- tissue histology and/or fluid cytology. Molecular testing may also be used
- Most Brain & CNS tumours are not considered stageable, but certain specified morphologies of tumour require a Chang stage
- Some brain / CNS tumours are treated with surgery. This may be to debulk the tumour, but complete removal can be difficult due to the risk of damage to surrounding tissue
- Radiotherapy works well with some tumours and may be applied in the form of high dose Radiosurgery
- The range of Chemotherapy available is limited due to the effect of the blood-brain barrier. Some Chemotherapy drugs can be inserted directly into the brain in the form of a gel wafer
- For advanced tumours no active treatment is offered, only Palliative treatment to improve the patient's symptoms

Where the tumour is advanced it might be the case that no active treatment is offered to the patient. Palliative treatment may be offered instead to control symptoms.

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

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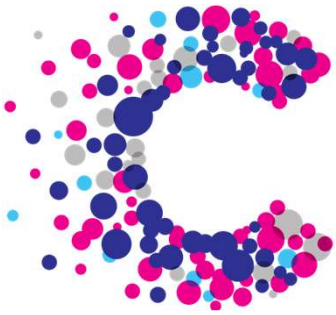

CANCER RESEARCH UK

We'd like to thank Cancer Research UK for the use of their images within this training module.

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If you have any questions on the information contained within this module or about COSD in general, do please feel free to email your regional Data Liaison Manager