



CNS tumours

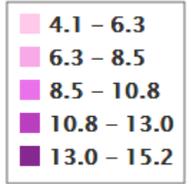
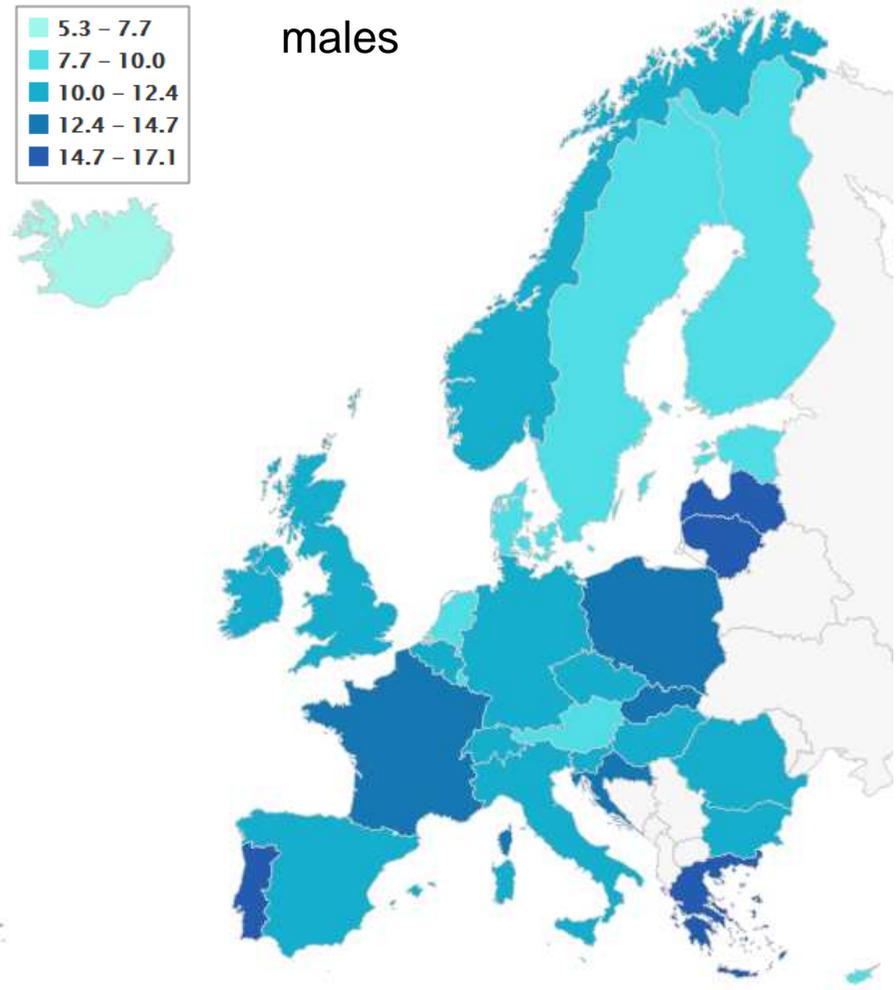
Coding issues

Otto Visser
June 2019

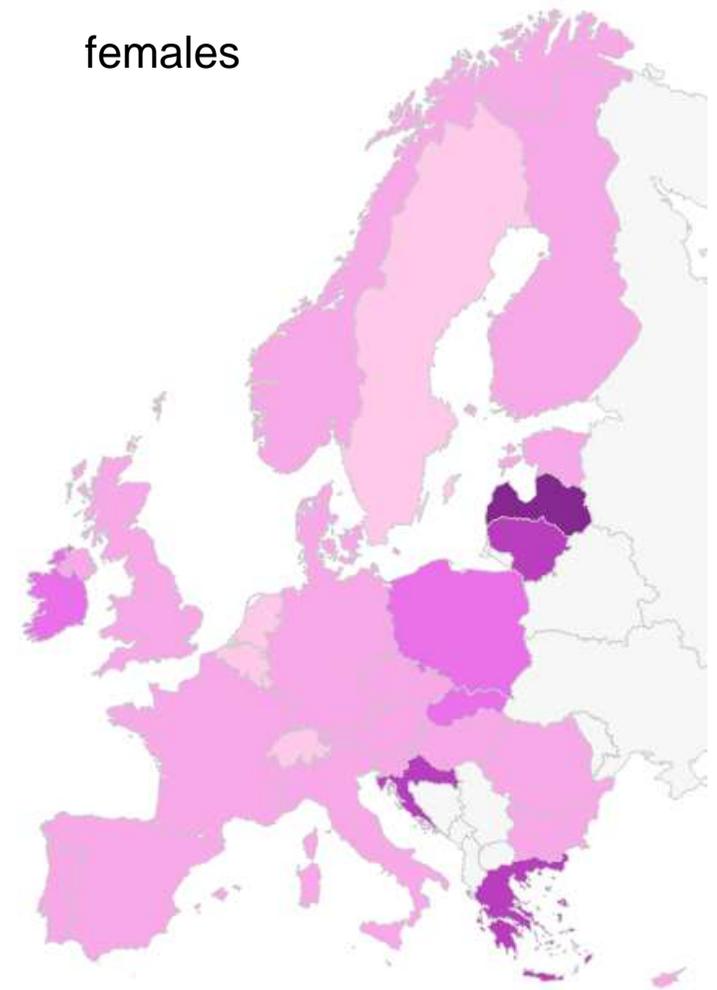
Incidence of CNS tumours in Europe in 2018



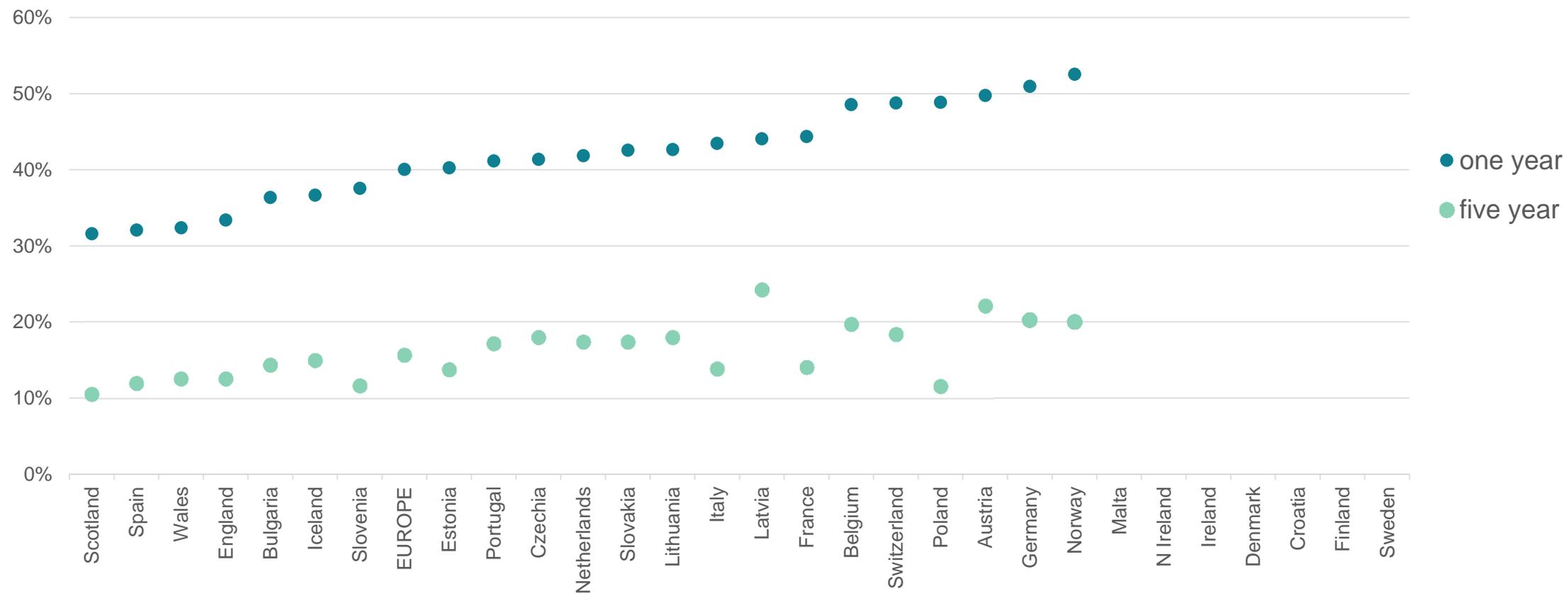
males



females

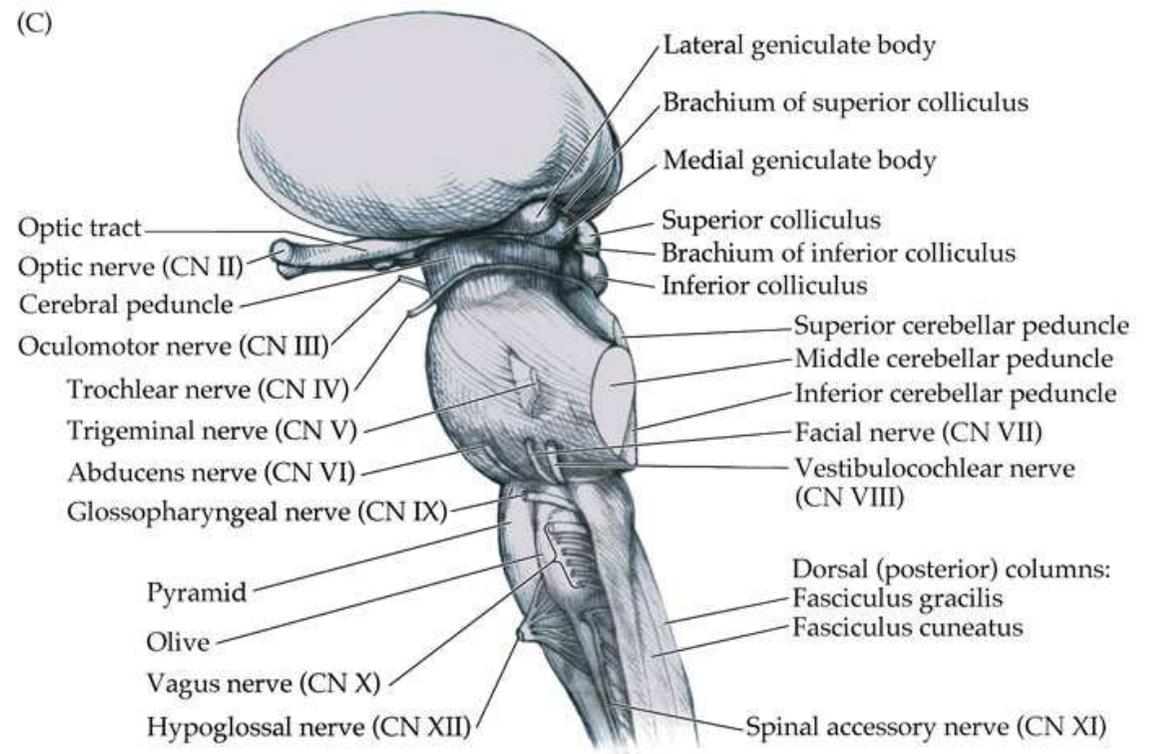


Relative survival of brain tumours(2000-2007)



Cranial nerves

- Olfactory nerve (I) C72.2
- Optic nerve (II) C72.3
- Acoustic nerve (VIII) C72.4
- Other cranial nerves C72.5



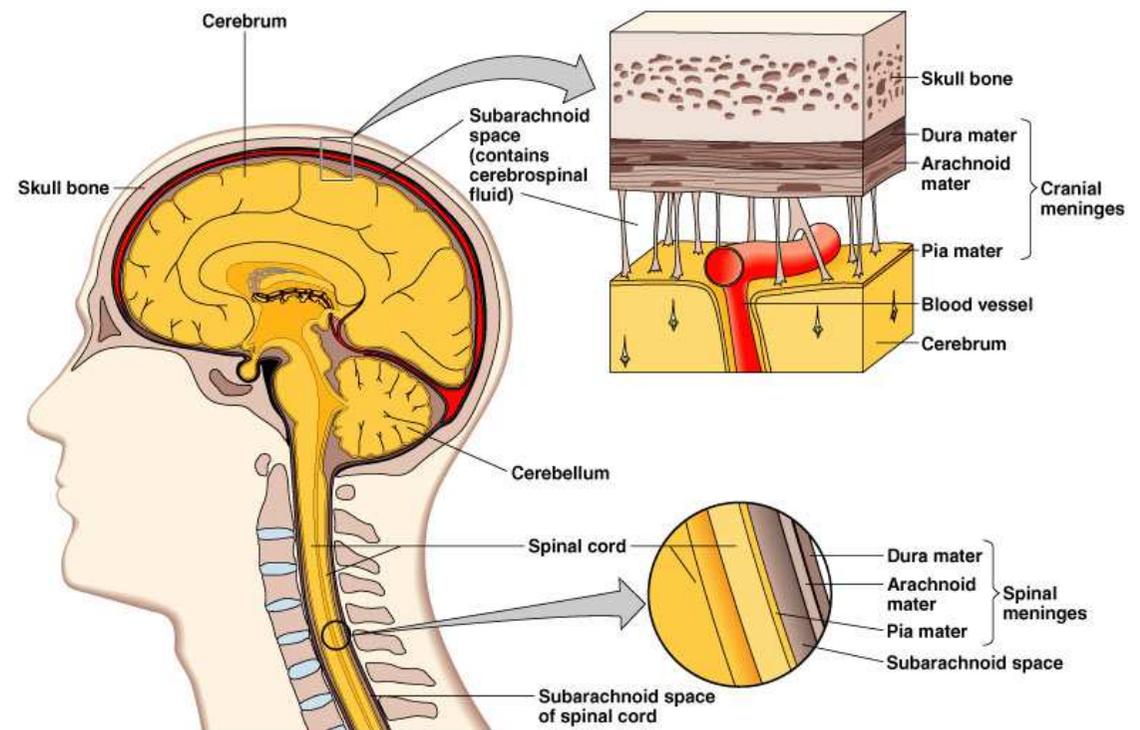
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Tumours of the cranial nerves

- Pilocytic astrocytoma (optic nerve)
 - in children
 - may be bilateral or in the chiasma
- Schwannoma (mostly acoustic nerve, but also in other cranial nerves)
 - vestibular schwannoma, acoustic neurinoma
 - benign in the vast majority of cases (M9560/0)
 - malignancy extremely rare (MPNST=M9540/3)
 - often the diagnosis is made on imaging only
 - may be bilateral

Meninges

- Cerebral meninges C70.0
- Spinal meninges C70.1



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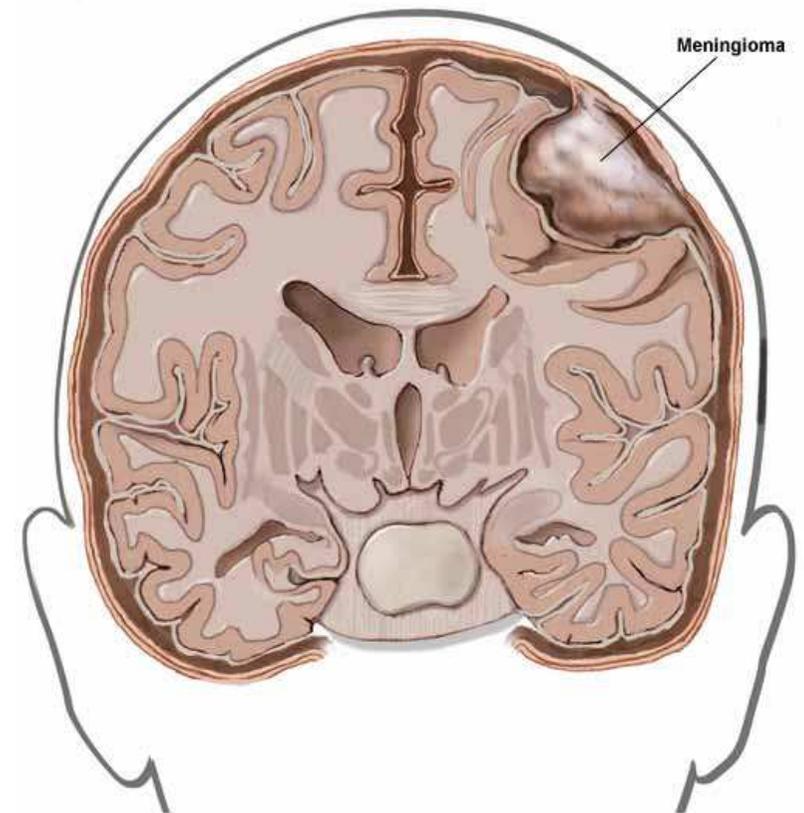
Tumours of the meninges

- Meningioma
 - Mostly benign (9530/0 – 9537/0)
 - Atypical (9538/1)
 - Rarely malignant (malignant meningioma = 9530/3 or meningeal sarcomatosis = 9539/3)
- Hemangiopericytoma
- Melanoma
 - Solitary (8720/3) or diffuse (8728/0, 8728/1, 8728/3)

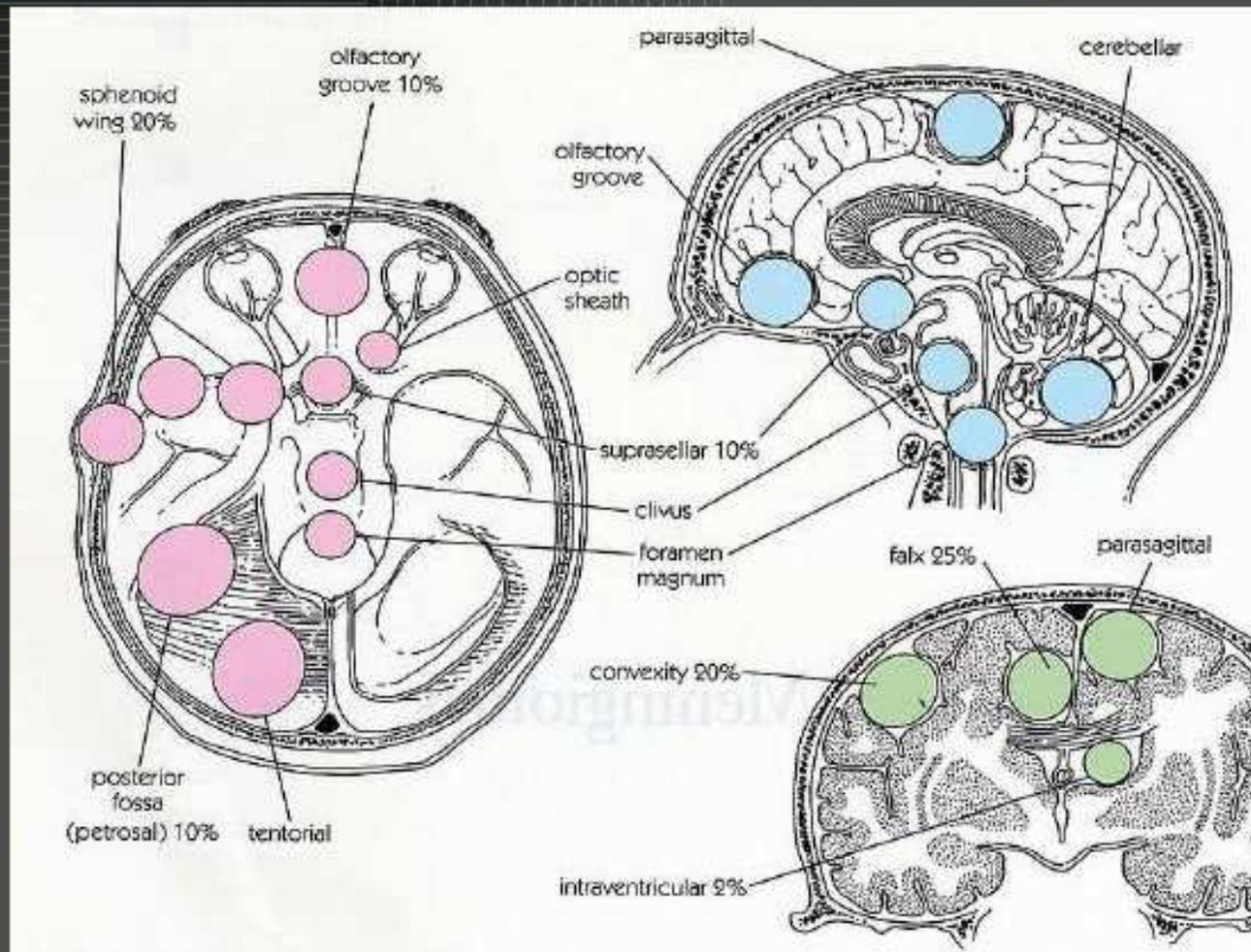
Tumours of the meninges

A meningioma of the CNS should always be coded on C70!

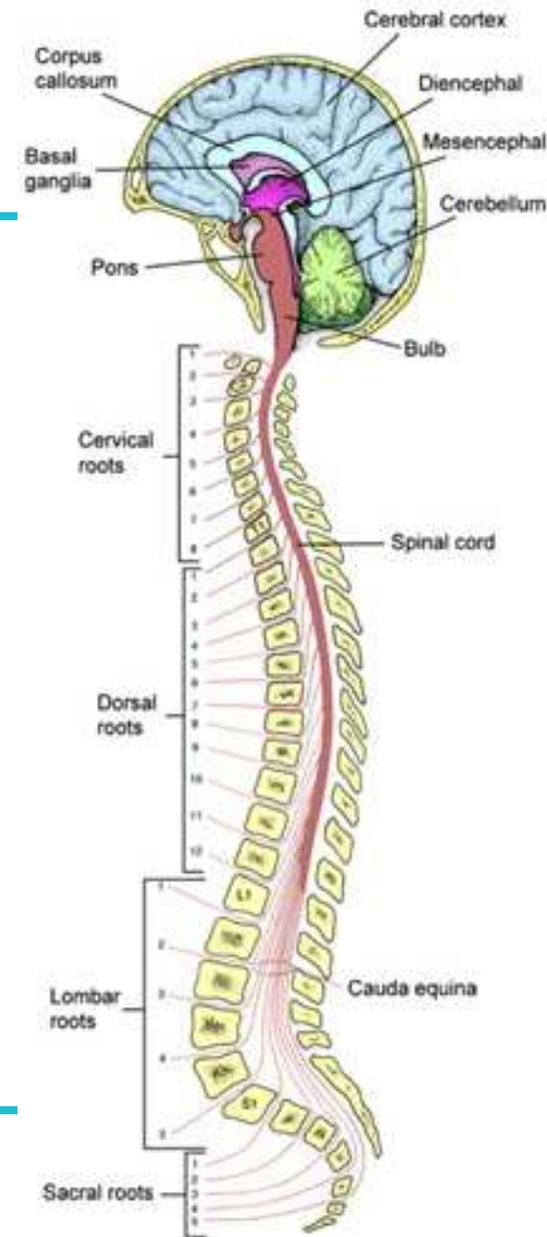
- 'Meningioma of the brain' = C70.0
- 'Meningioma of the spinal cord' = C70.1



Location



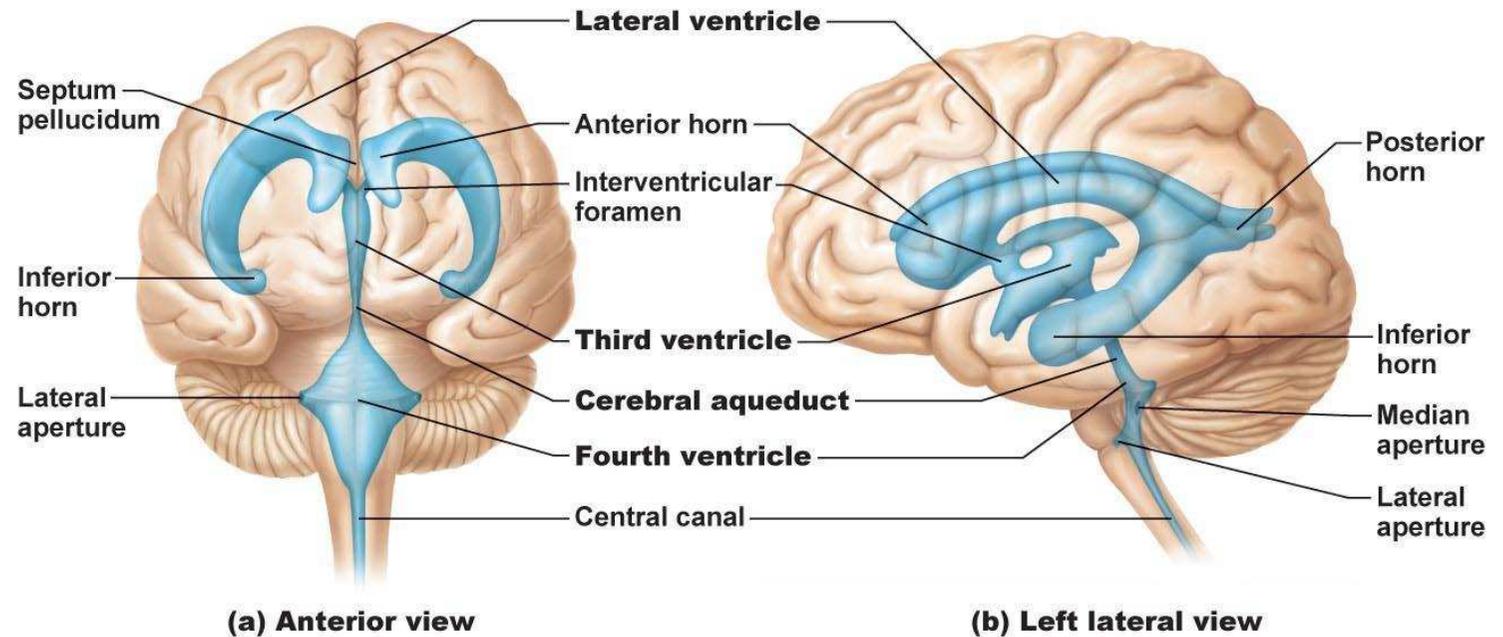
Brain and spinal cord



- Cortex
 - Frontal lobe (C71.1)
 - Temporal lobe (C71.2)
 - Parietal lobe (C71.3)
 - Occipital lobe (C71.4)
- Basal ganglia (C71.0)
- Cerebellum (C71.6)
- Brain stem (C71.7)
- Ventricles (C71.5)
- Spinal cord/cauda equina (C72.0/C72.1)

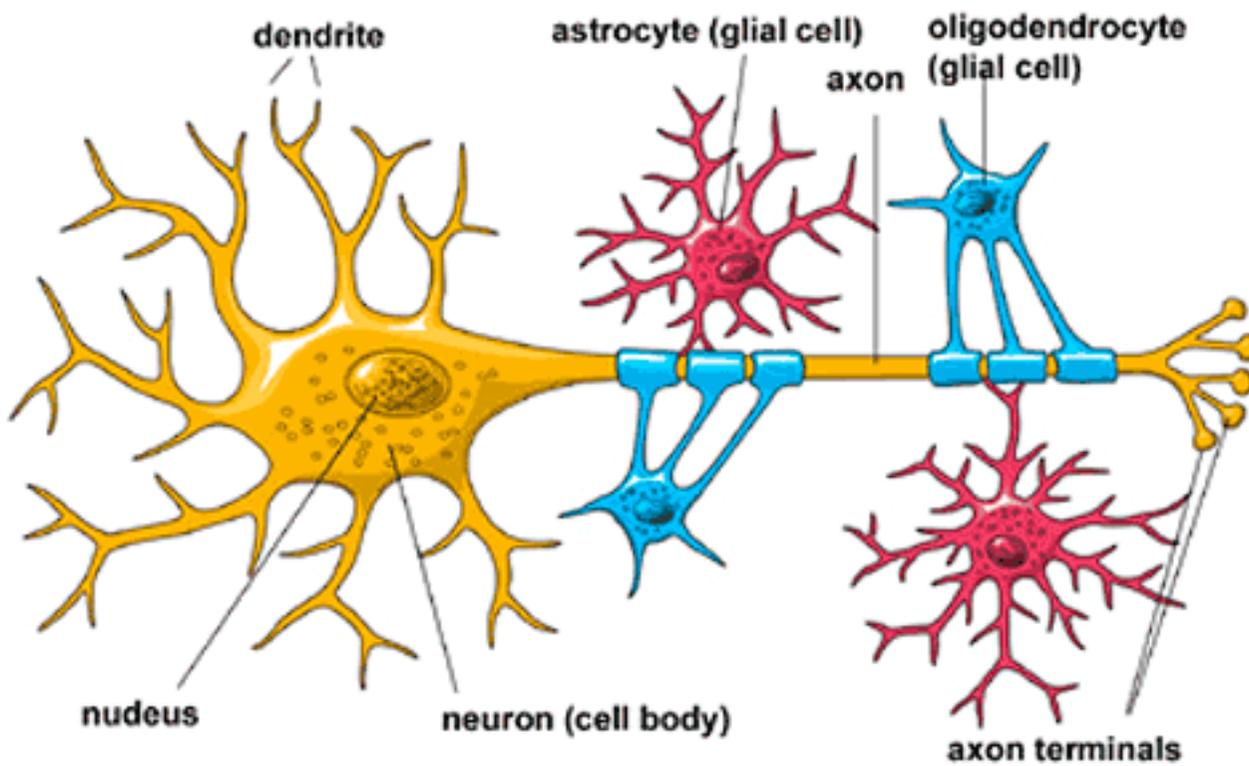
The ventricles of the brain

- The ventricles are filled with fluid; the only anatomical structure in the ventricles is the choroid plexus



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Brain cells



Neuro-epithelial tumours of the brain and spinal cord

- Gliomas (glioma='tumour of glial cell')
 - Astrocytic tumours
 - Oligodendroglial tumours
 - Oligo-astrocytic tumours
 - Ependymal tumours
 - Choroid plexus tumours
- Neuronal and mixed neuronal-glial tumours
- Tumours of the epiphysis
- Embryonal tumours

WHO grade

The grade aims to predict the biological behaviour of the tumour

- Grade I: tumour with low proliferation and potential cure after resection
- Grade II: infiltrative tumour with low proliferation but with risk of recurrence after resection
- Grade III: histological malignant tumour (nuclear atypia & many mitoses) which requires (adjuvant) Rt and/or Ct after resection
- Grade IV: histological malignant tumour with necrosis and fast progression with fatal outcome

Transformation from a lower to a higher grade can occur (as in HM)

Grading in CNS tumours differs from other cancers

- low grade = WHO grade 2
- high grade = WHO grade 3
- 'anaplastic' = WHO grade 3

- *a specific WHO grade overrules a descriptive term*

Site of the tumour

- Astrocytoma: 98% cerebral
- Oligodendroglioma: 99% cerebral
- Ependymoma: 50% cerebral (the wall of the ventricles), 50% spinal (the spinal canal)
 - Subependymoma & anaplastic ependymoma mostly cerebral
 - Myxopapillary ependymoma mostly spinal (cauda equina/filum terminale)
- Choroid plexus tumor: always cerebral (ventricle)

New morphology codes in 1st revision of ICD-O-3

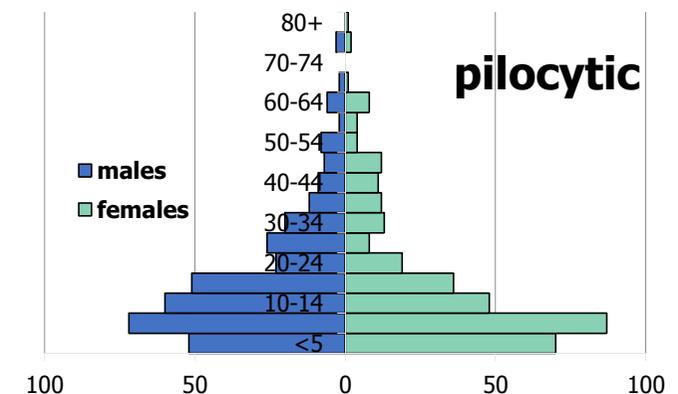
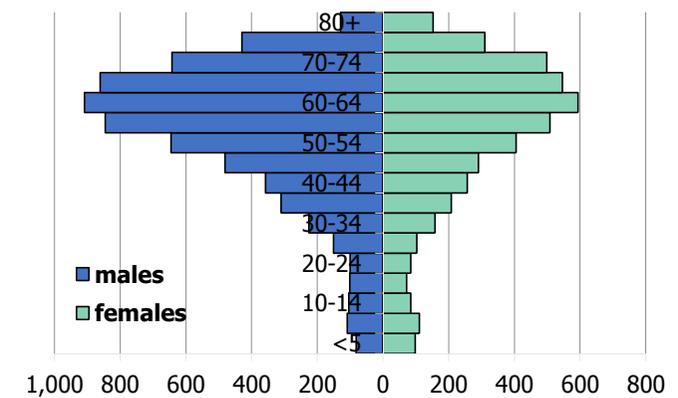
Code	Term
9395/3	Papillary tumor of the pineal region
9425/3	Pilomyxoid astrocytoma
9431/1	Angiocentric glioma
9432/1	Pituicytoma
9509/1	Papillary (Rosette-forming) glioneuronal tumor

New morphology codes in 2nd revision of ICD-O-3

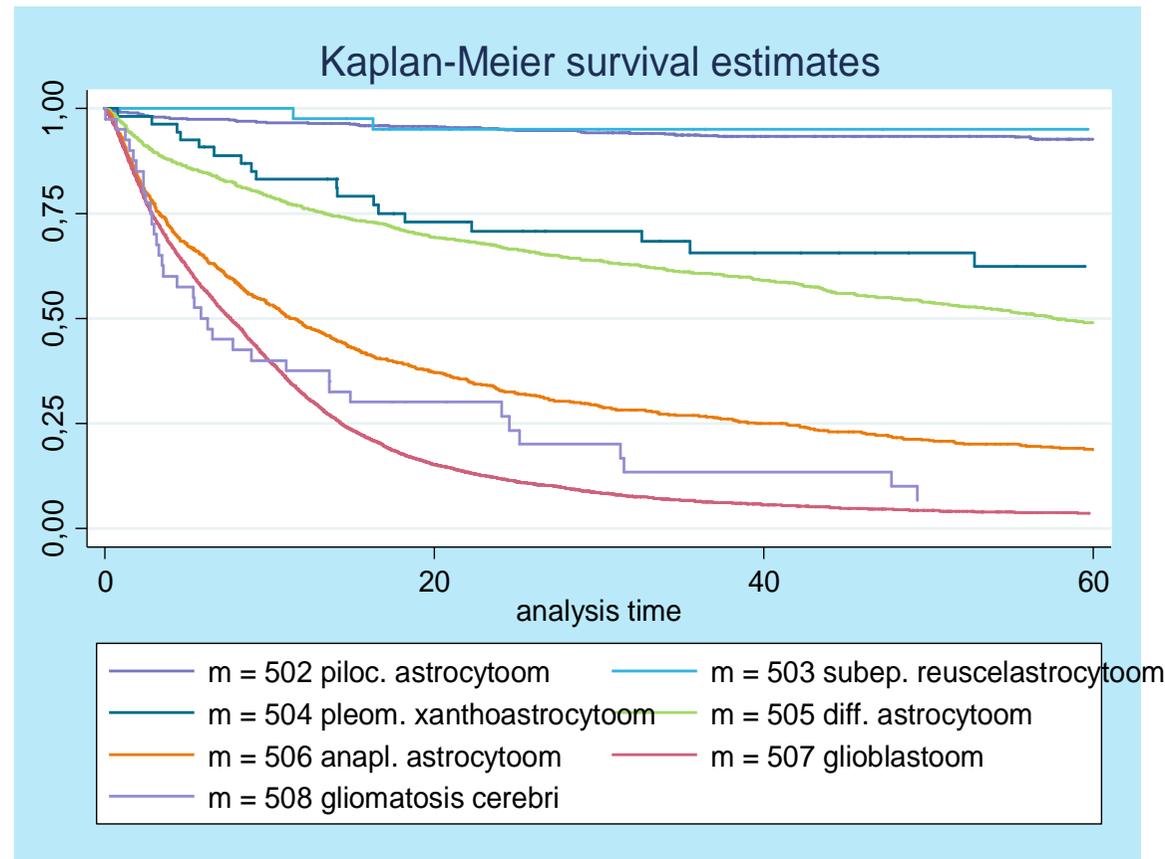
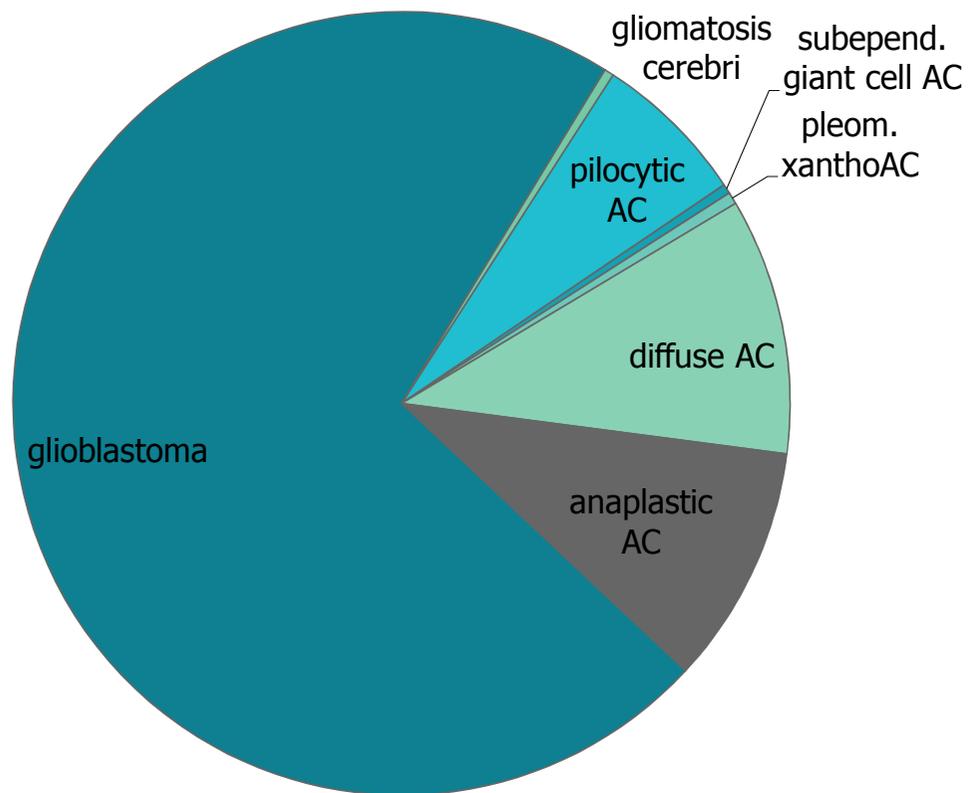
Code	Term
9385/3	Diffuse midline glioma, H3 K27M-mutant (C71._)
9396/3	Ependymoma, RELA fusion-positive (C71._)
9445/3	Glioblastoma, IDH-mutant (C71._)
9475/3	Medulloblastoma, WNT-activated, classic
9476/3	Medulloblastoma, SHH-activated and TP53 mutant
9477/3	Medulloblastoma, non-WNT/non-SHH
9478/3	Embryonal tumor with multilayered rosettes with C19MC alteration (C71._)
9542/3	Epithelioid MPNST

Astrocytoma: classification and age distribution

type	Morphology	WHO grade
Pilocytic astrocytoma subtype: pilomyxoid astrocytoma (9425/3, grade II)	9421/1	I
Subependymal giant cell astrocytoma	9384/1	I
Pleomorphic xanthoastrocytoma	9424/3	II
Diffuse astrocytoma subtypes: fibrillary, gemistocytic, protoplasmic	9400/3 9420/3, 9411/3, 9410/3	II
Anaplastic astrocytoma	9401/3	III
Glioblastoma subtypes: giant cell glioblastoma, gliosarcoma	9440/3 9441/3, 9942/3	IV
Gliomatosis cerebri	9381/3	mostly III

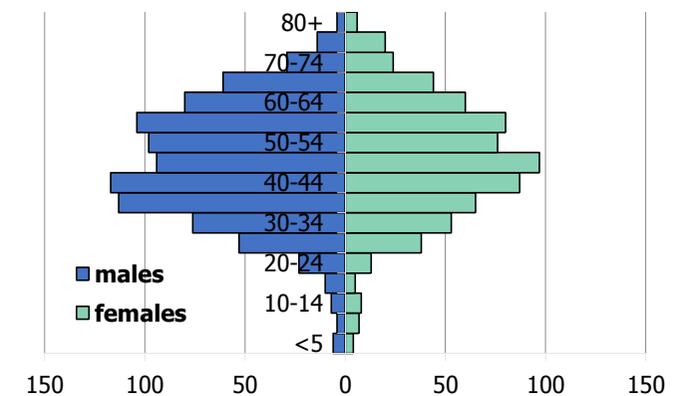


Astrocytoma

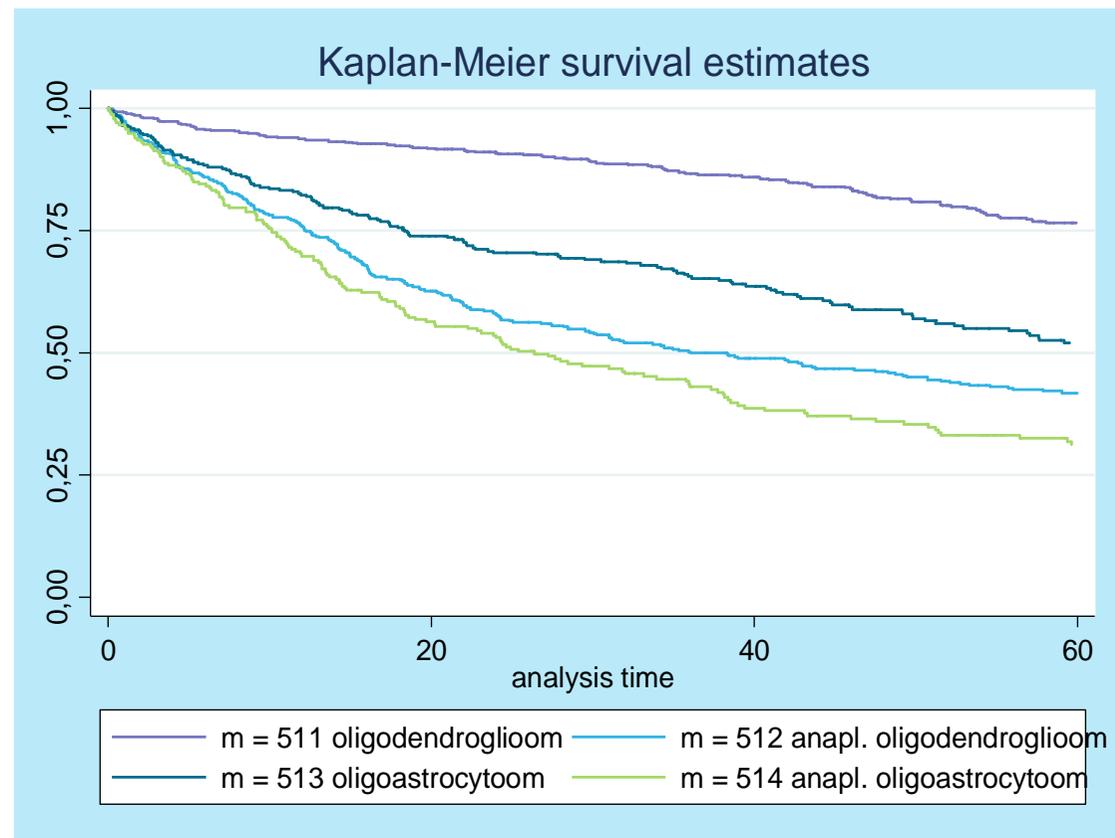
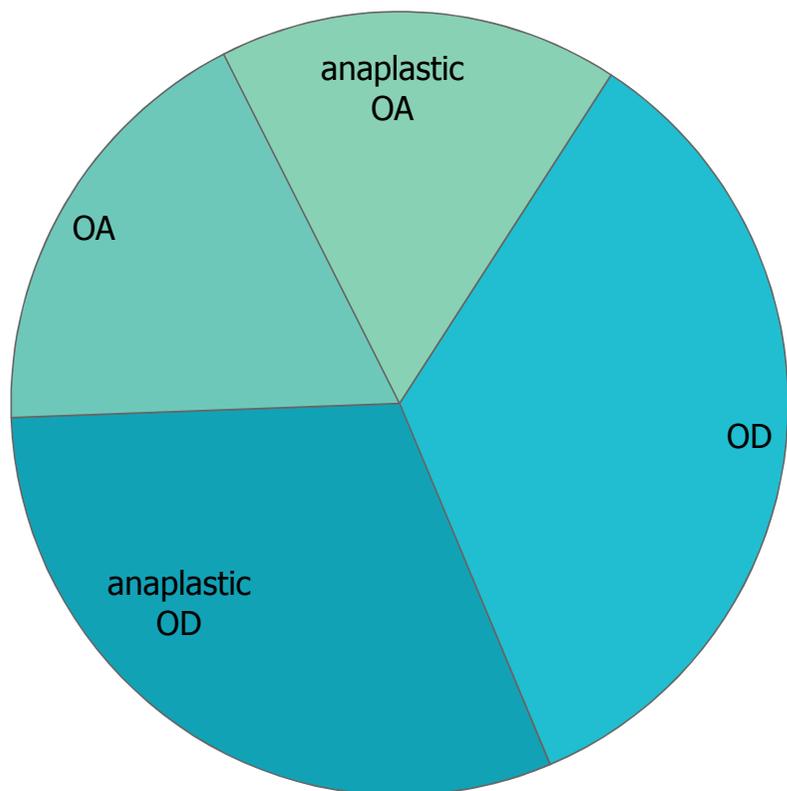


Oligodendroglioma: classification and age distribution

type	Morphology	WHO grade
Oligodendroglioma	9450/3	II
Anaplastic oligodendroglioma	9451/3	III
Oligoastrocytoma	9382/3	II
Anaplastic oligoastrocytoma	9382/3	III

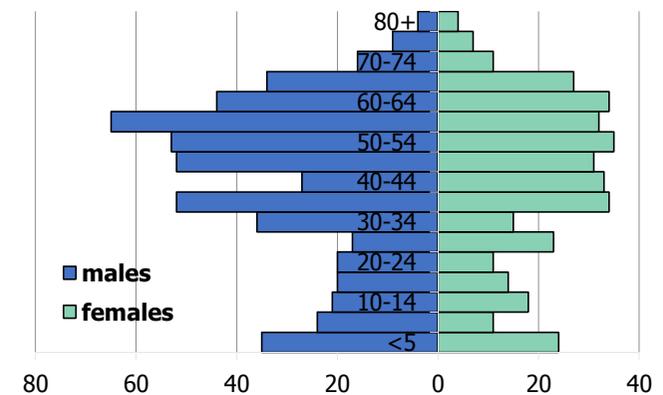


Oligodendroglioma

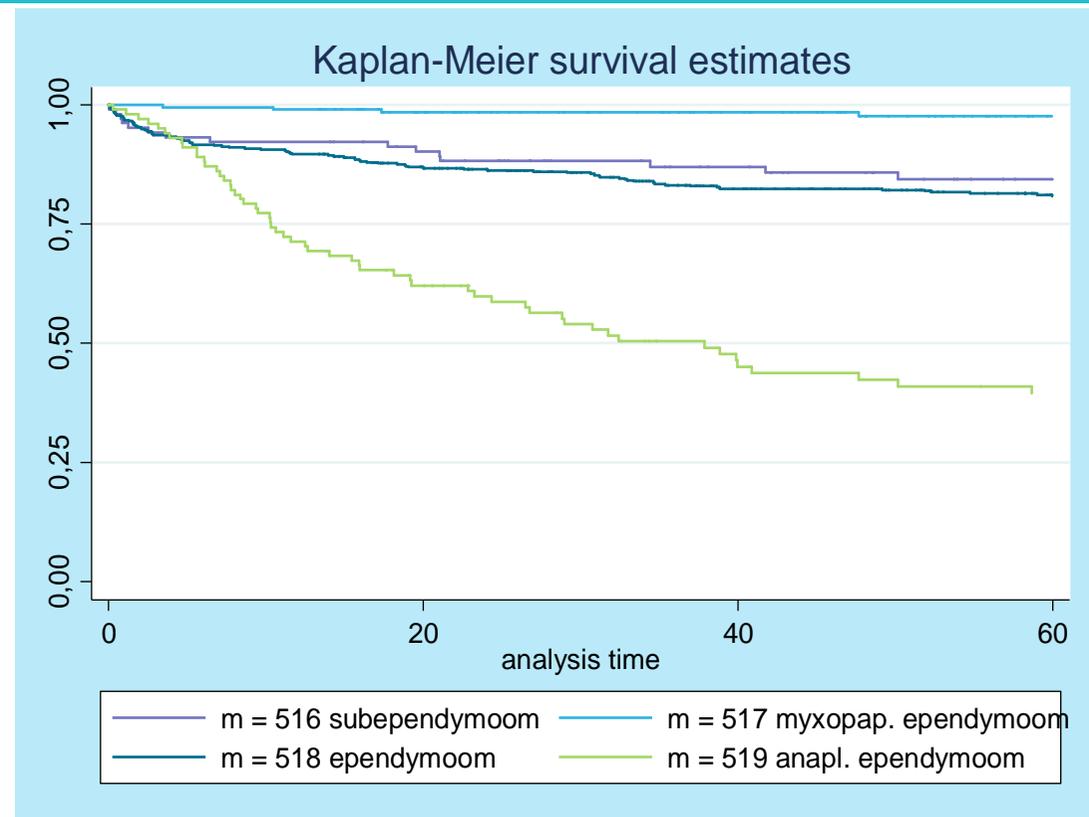
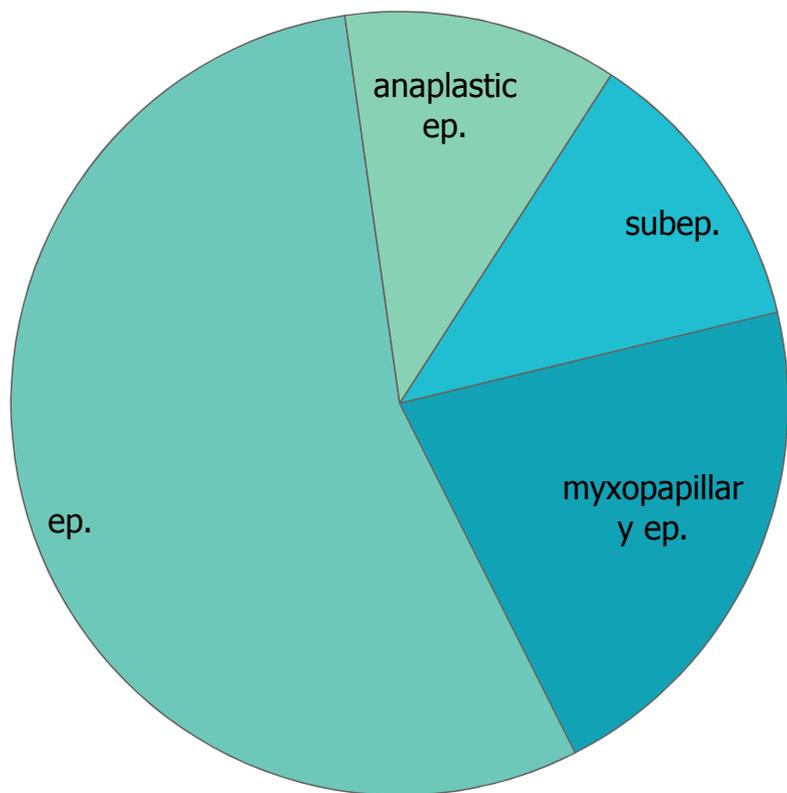


Ependymoma: classification and age distribution

type	Morphology	WHO grade
Subependymoma	9383/1	I
Myxopapillary ependymoma	9394/1	I
Ependymoma (subtypes: cellulai, papillary (9393/3), clear cell, tancytic)	9391/3	II
Anaplastic ependymoma	9392/3	III

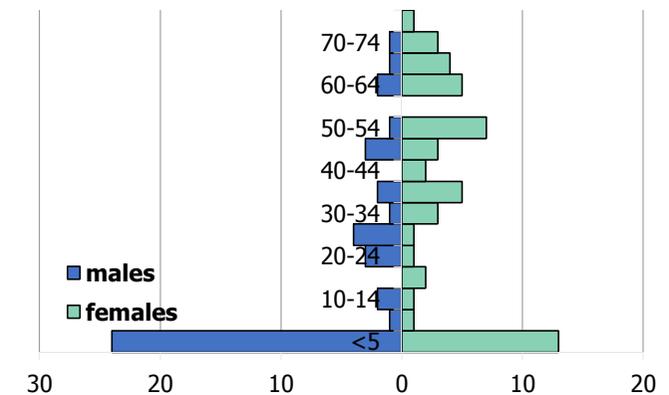


Ependymoma

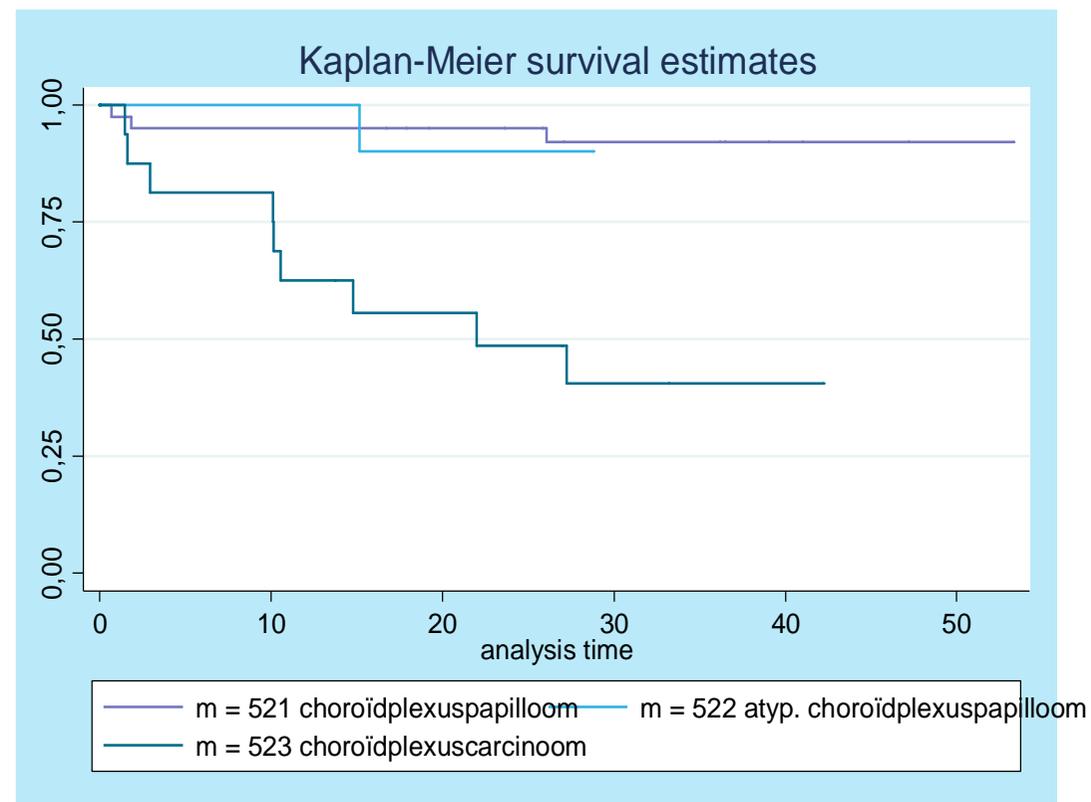
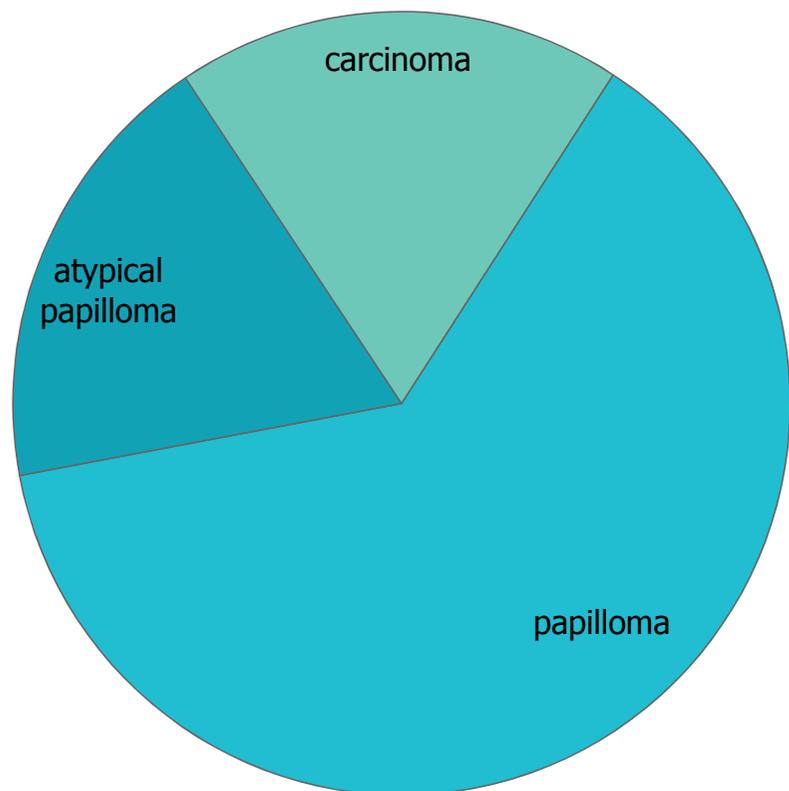


Choroid plexus tumours: classification and age distribution

type	Morphology	WHO grade
Choroid plexus papilloma	9390/0	I
Choroid plexus papilloma, atypical	9390/1	II
Choroid plexus carcinoma	9390/3	III

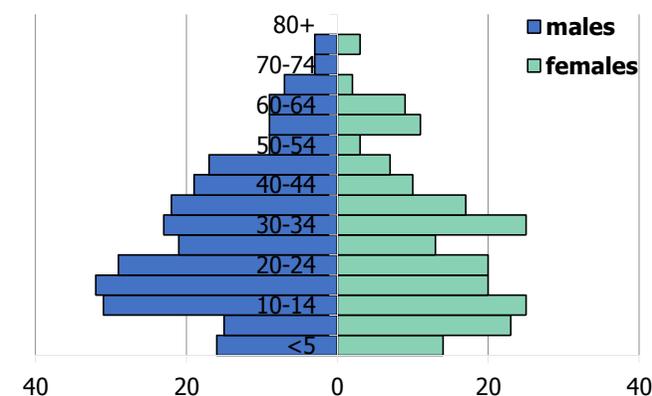


Choroid plexus tumours



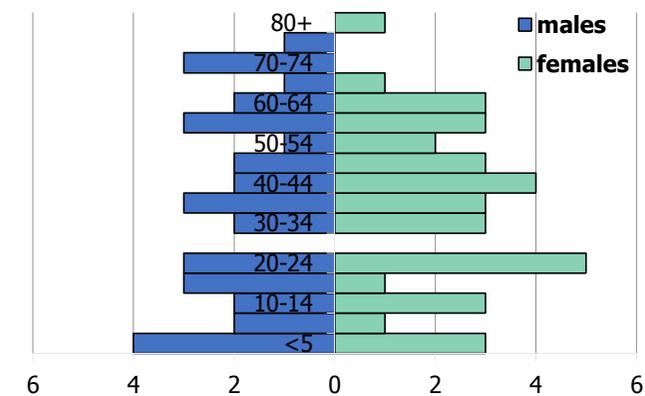
Neuronal & mixed neuronal-glial tumours: classification and age distribution

type	Morphology	WHO grade
Dysplastic gangliocytoma of the cerebellum	9493/0	I
Desmoplastic infantile astrocytoma/ganglioglioma	9412/1	I
Dysembryoplastic neuroepithelial tumour	9413/0	I
Gangliocytoma	9492/0	I
Ganglioglioma	9505/1	I (or II)
Anaplastic ganglioglioma	9505/3	III
Neurocytoma (central, extraventricular, cerebellar)	9506/1	I or II
Papillary glioneural tumour	9509/1	I
Spinal paraganglioma	8680/1	I

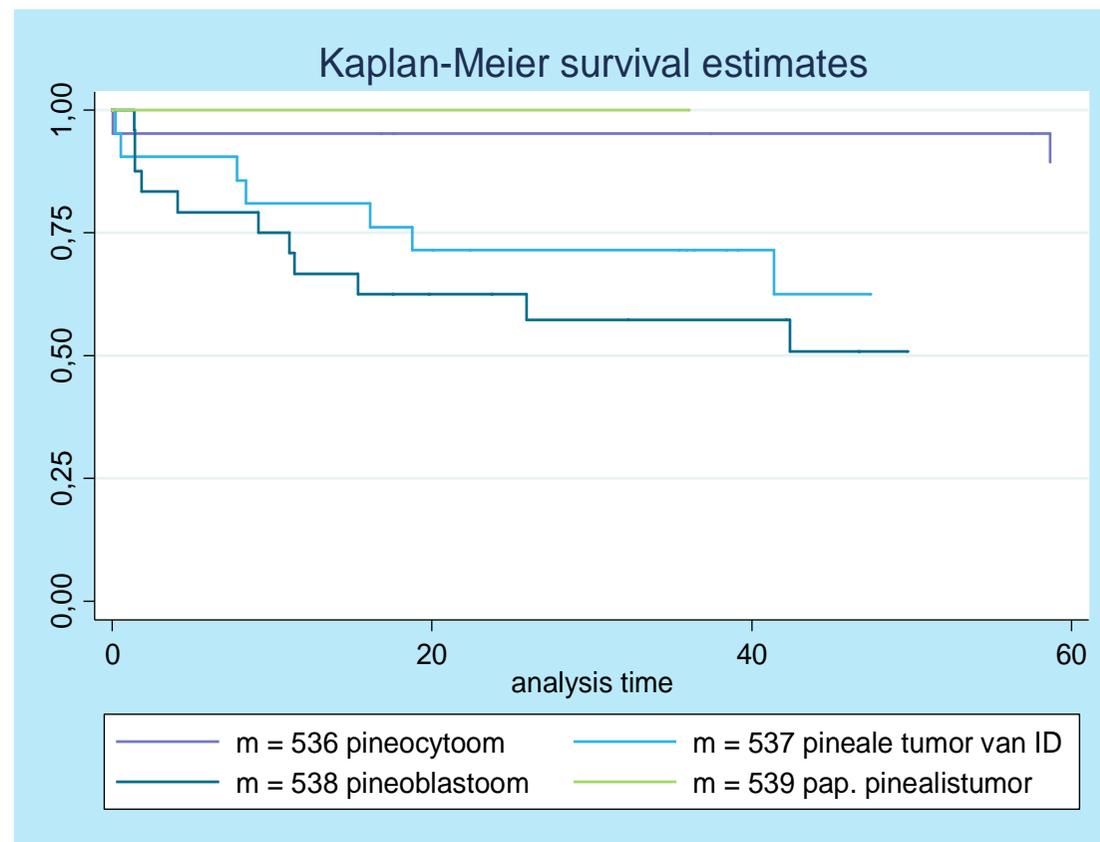
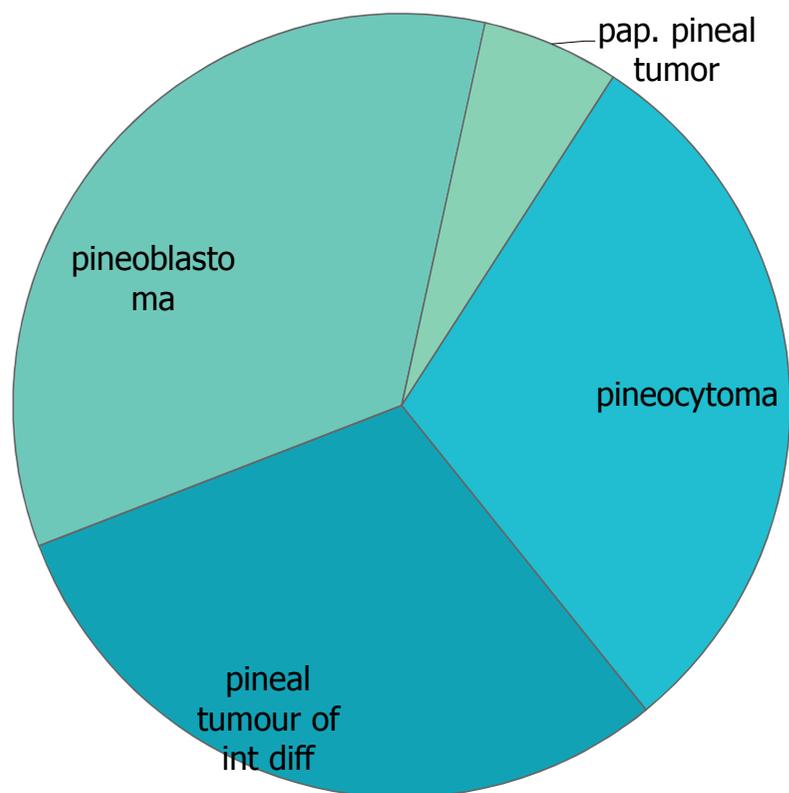


Tumours of the pineal region: classification and age distribution

type	Morphology	WHO grade
Pineocytoma	9361/1	I (was II)
Pineal parenchymal tumour of intermediate differentiation	9362/3	II of III (was III of IV)
Pineoblastoma	9362/3	IV
Papillary tumour van de pinealis region	9395/3	II of III

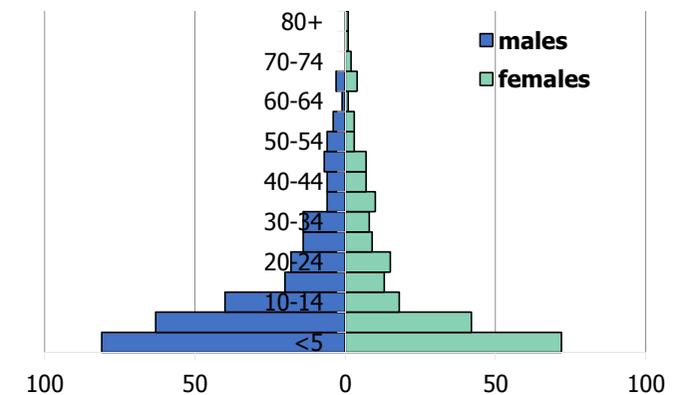


Tumours of the pineal region

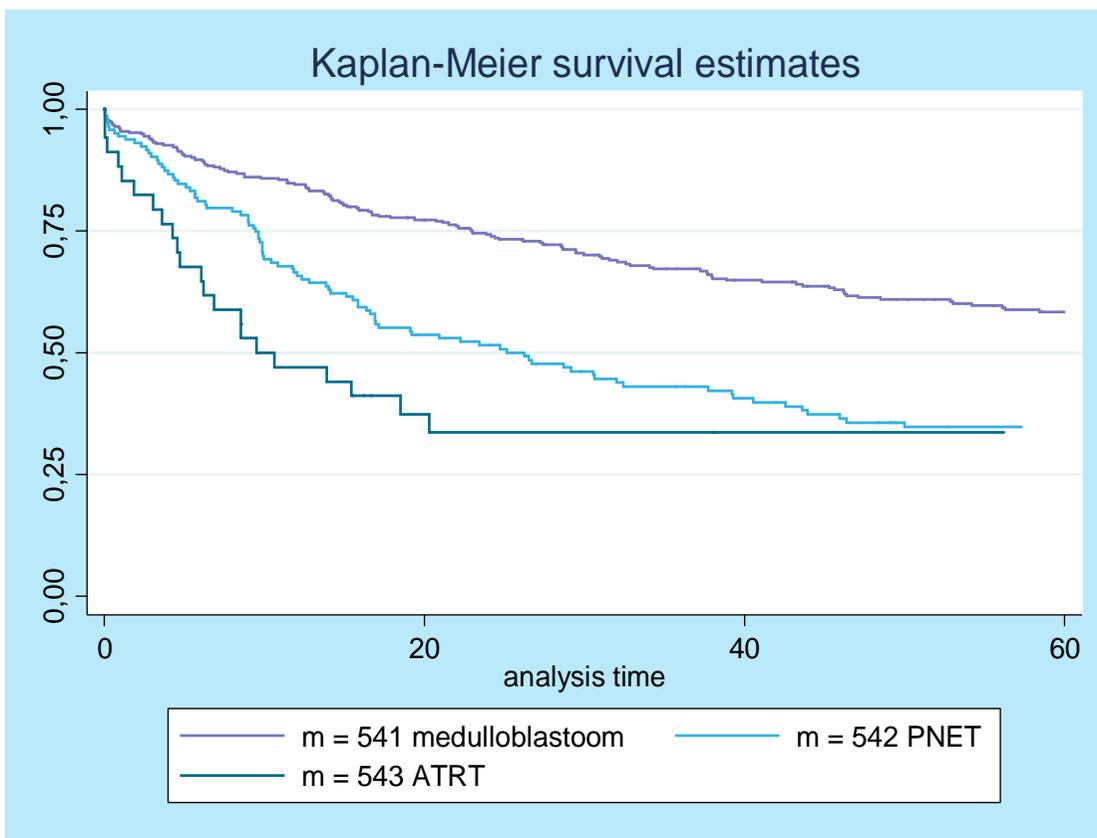
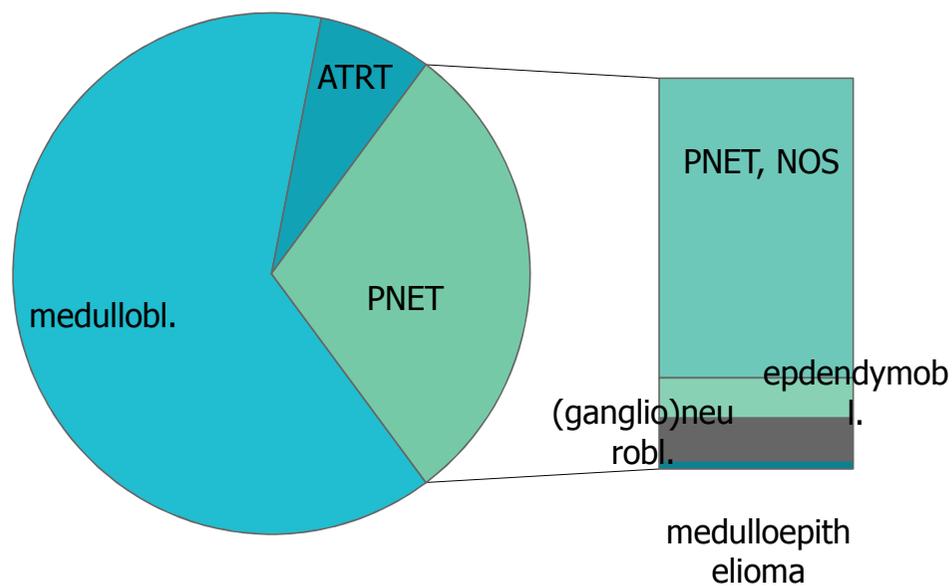


Embryonal tumours: classification and age distribution

type	Morphology	WHO grade
Medulloblastoma (subtypes: desmoplastic/nodular, anaplastic/large cell)	9470/3 (9471/3, 9474/3)	IV
PNET of the CNS (subtypes: neuroblastoma, ganglioneuroblastoma, medullo-epithelioma, ependymoblastoma)	9473/3 (9500/3, 9490/3, 9501/3, 9392/3)	IV
Atypical teratoid / rhabdoid tumour	9508/3	IV



Embryonal tumours





EXERCISES



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