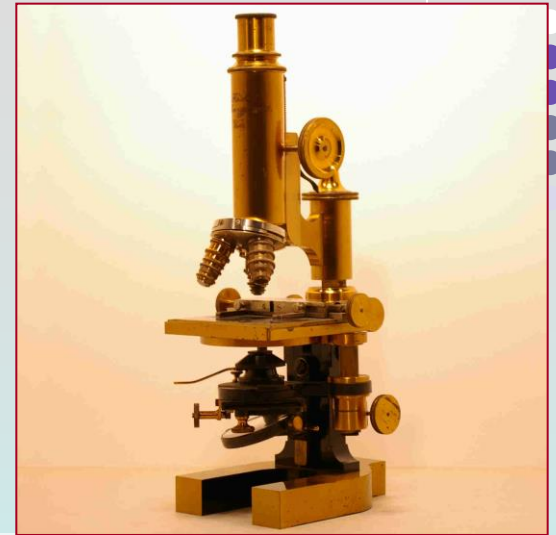




*Associazione Italiana Registri Tumori*  
*Corso autunnale*  
*Camerino, 11-13 settembre 2013*



## **I tumori dell'encefalo e del Sistema Nervoso Centrale**

*Stefano Ferretti*  
*Registro Tumori Area Vasta Emilia Centrale*

«benigno»

«maligno»

differenziazione

prognosi

base diagnosi

inquadramento e peculiarità

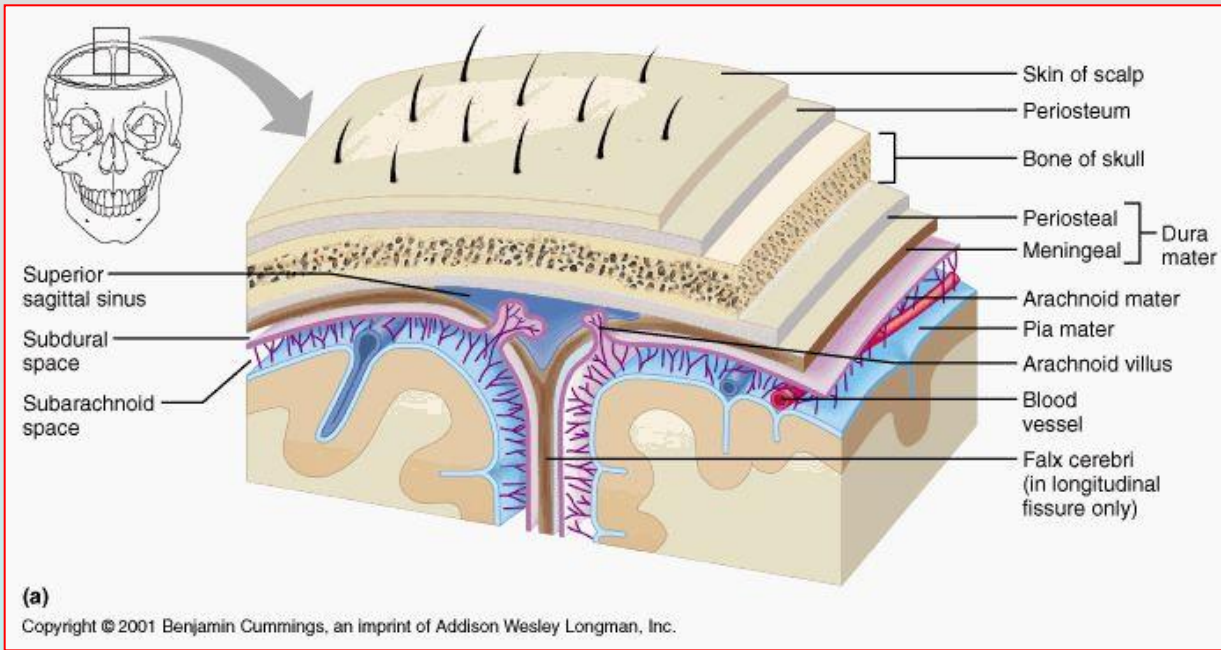
infiltrazione (?)

attività proliferativa  
morfologia

istologia  
grading  
topografia

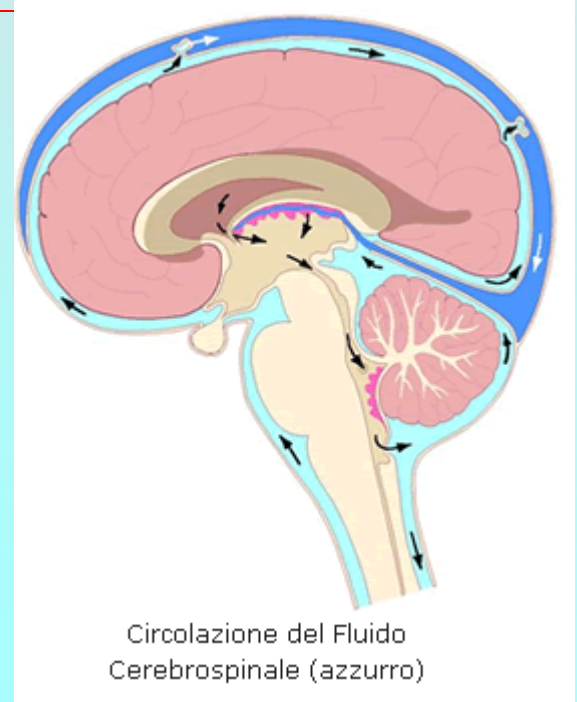
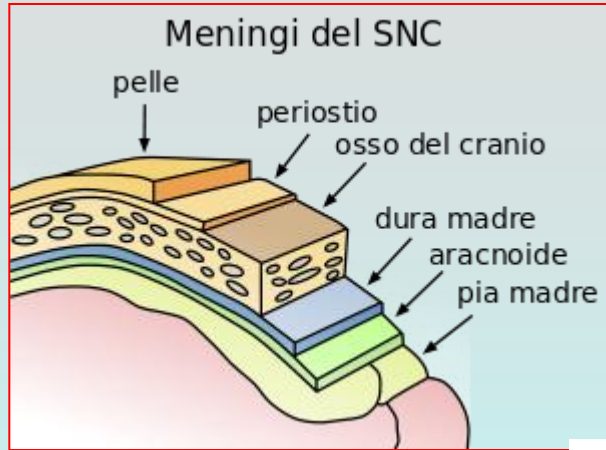
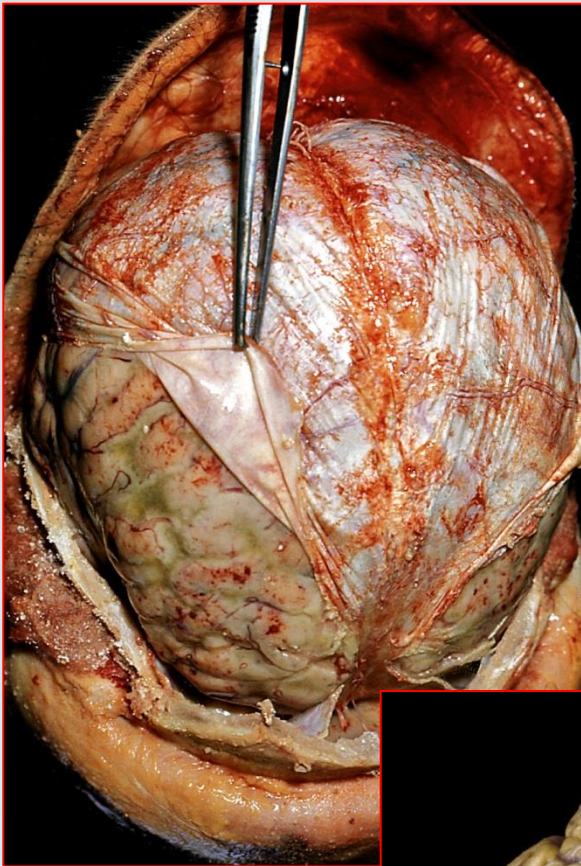
anatomia patologica  
imaging





# Meningi

# anatomia delle meningi



# neoplasie delle meningi



## tumori delle cellule meningoteliali

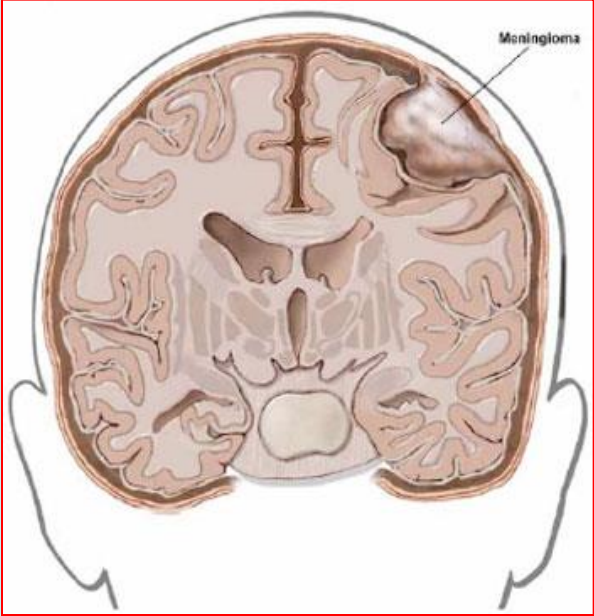
Acta Neuropathol (2007) 114:97–109  
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REVIEW

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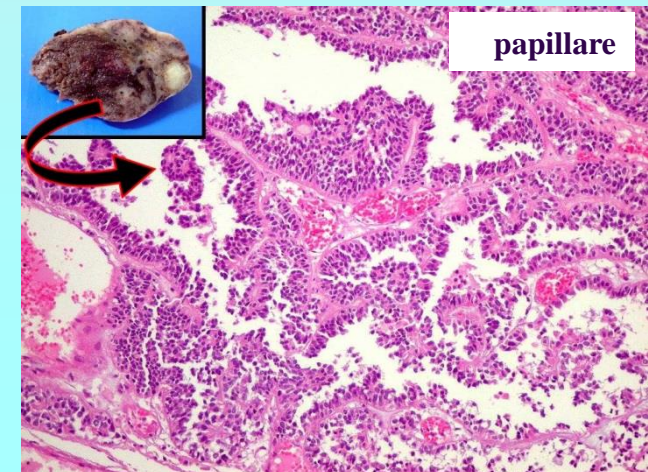
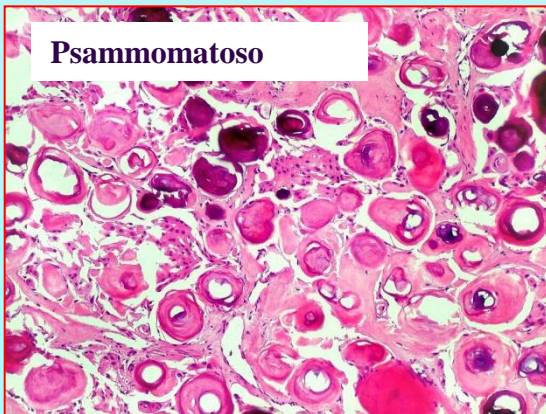
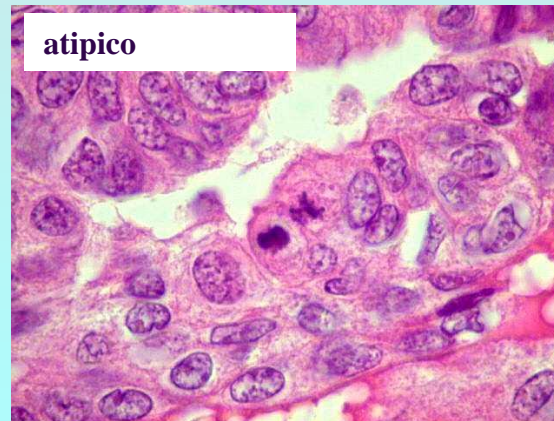
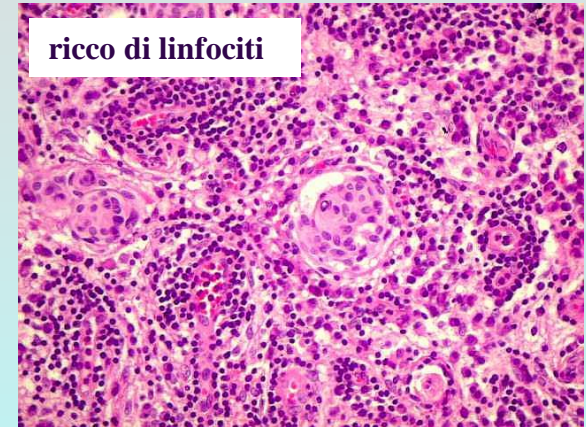
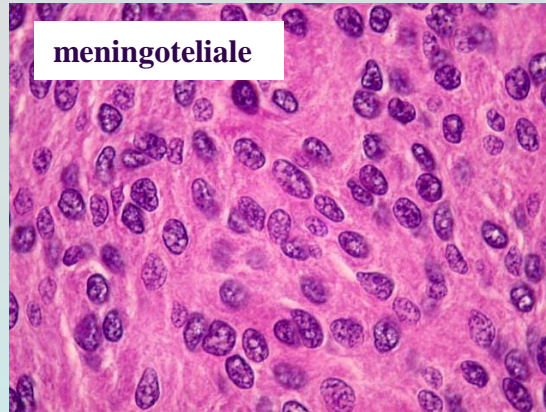
• NAS	9530/0
• Meningoteliale	9531/0
• Fibroso	9532/0
• Transizionale	9537/0
• Psammomatoso	9533/0
• Angiomatoso	9534/0
• Microcistico	9530/0
• Secretorio	9530/0
• Ricco di linfociti	9530/0
• Metaplastico	9530/0
• Cordoide	9538/1
• A cell. chiare	9538/1
• Atipico	9539/1
• Papillare	9538/3
• Rabdoide	9538/3
• Anaplastico	9530/3



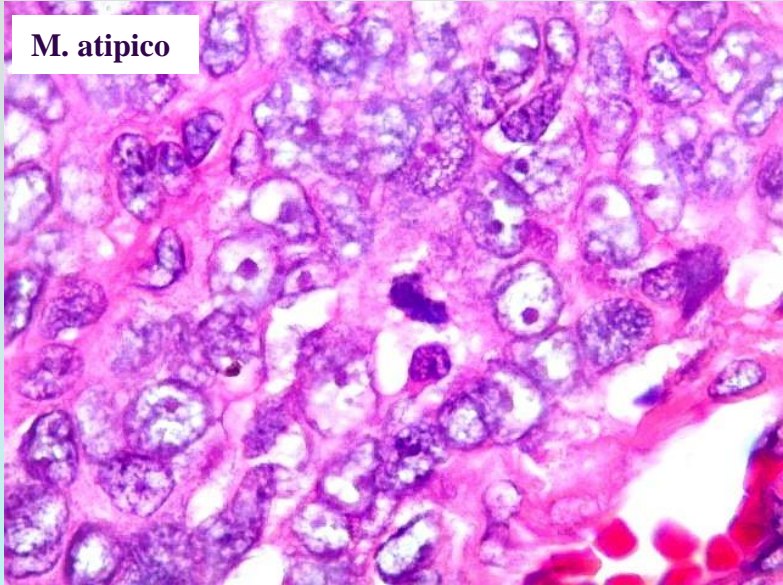
# neoplasie delle meningi



- NAS
- Meningoteliale
- Fibroso
- Transizionale
- Psammomatoso
- Angiomatoso
- Microcistico
- Secretorio
- Ricco di linfociti
- Metaplastico
- Cordoide
- A cell. chiare
- Atipico
- Papillare
- Rabdoide
- Anaplastico



# neoplasie delle meningi



M. atipico

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- ### Caratteri clinico-biologici
- Insorgenza in età adulta, forme «benigne»
  - Rischio da progressa rx terapia
  - Origini dai meningoteli dell'aracnoide
  - Spesso adesi alla dura
  - Ubiquitari, anche ventricolari (plessi)
  - Talora «infiltranti» teca ed encefalo
  - Aspetto morfologico e differenziativo estremamente polimorfo

Meningeal tumours	I	II	III	IV
Meningioma	•			
Atypical meningioma		•		
Anaplastic / malignant meningioma			•	
Haemangiopericytoma		•		
Anaplastic haemangiopericytoma			•	
Haemangioblastoma	•			



# neoplasie delle meningi

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## tumori mesenchimali

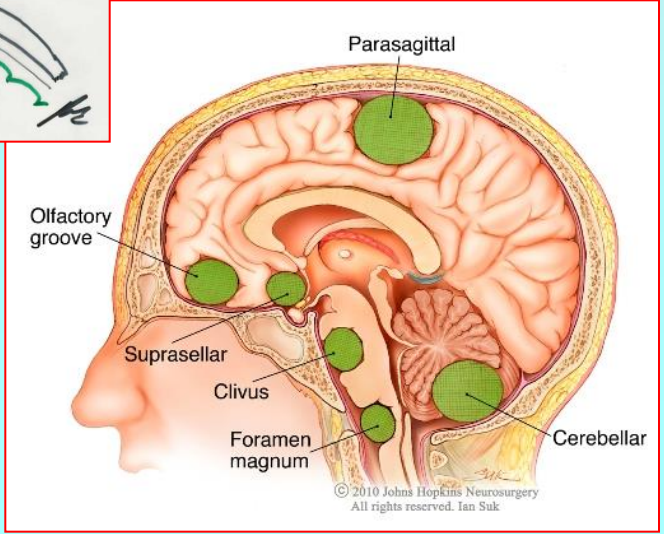
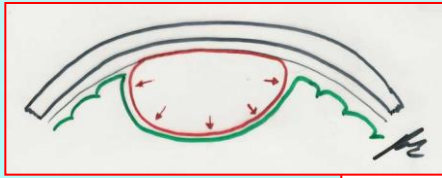
• Lipoma	8850/0
• Angiolipoma	8861/0
• Ibernoma	8880/0
• Liposarcoma	8850/3
• T. fibroso solitario	8815/0
• Fibrosarcoma	8810/3
• F. istiocitoma mal.	8830/3
• Leiomioma	8890/0
• Leiomiosarcoma	8890/3
• Rabdomioma	8900/0
• Rabdomiosarcoma	8900/3
• Condroma	9220/0
• Condrosarcoma	9220/3
• Osteoma	9180/0
• Osteosarcoma	9180/3
• Osteocondroma	9210/0
• Emangioma	9120/0
• Emagioendotelioma epit.	9133/1
• Emagiopericitoma	9150/1
• Emagiopericitoma anapl.	9150/3
• Angiosarcoma	9120/3
• S. di Kaposi	9140/3
• S. di Ewing + PNET	9364/3

## tumori melanocitari primitivi

• Melanocitosi diffusa	8728/0
• Melanocitoma	8728/1
• Melanoma maligno	8720/3
• Melanomatosi meningea	9140/3

## altre neoplasie meningee

• Emangioblastoma	9160/1
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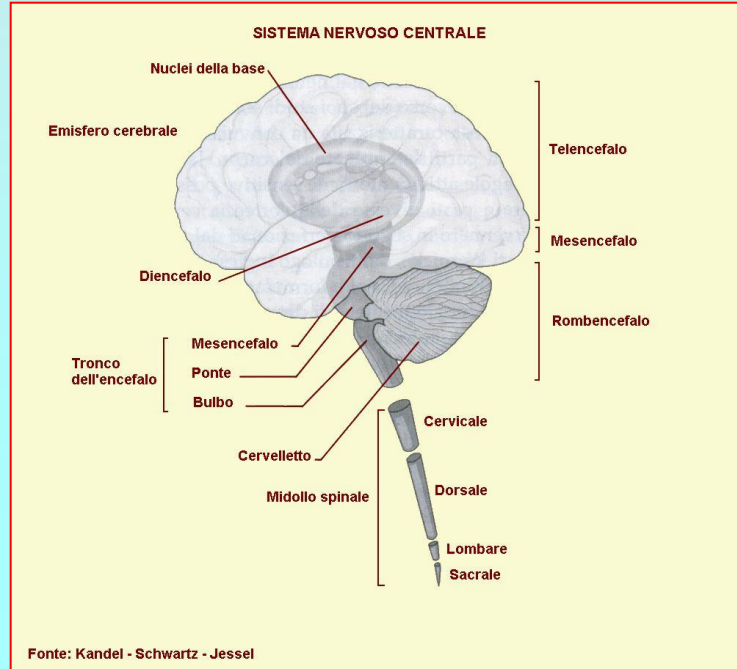
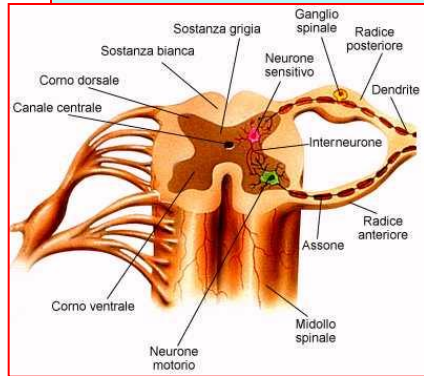
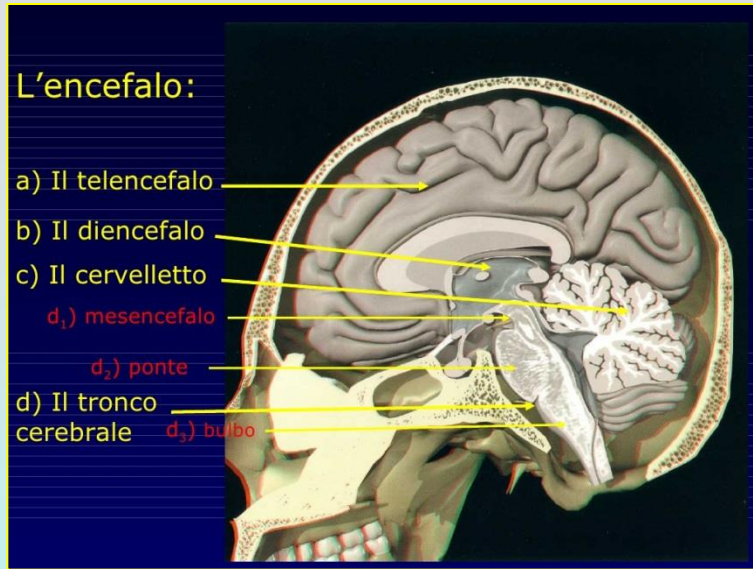
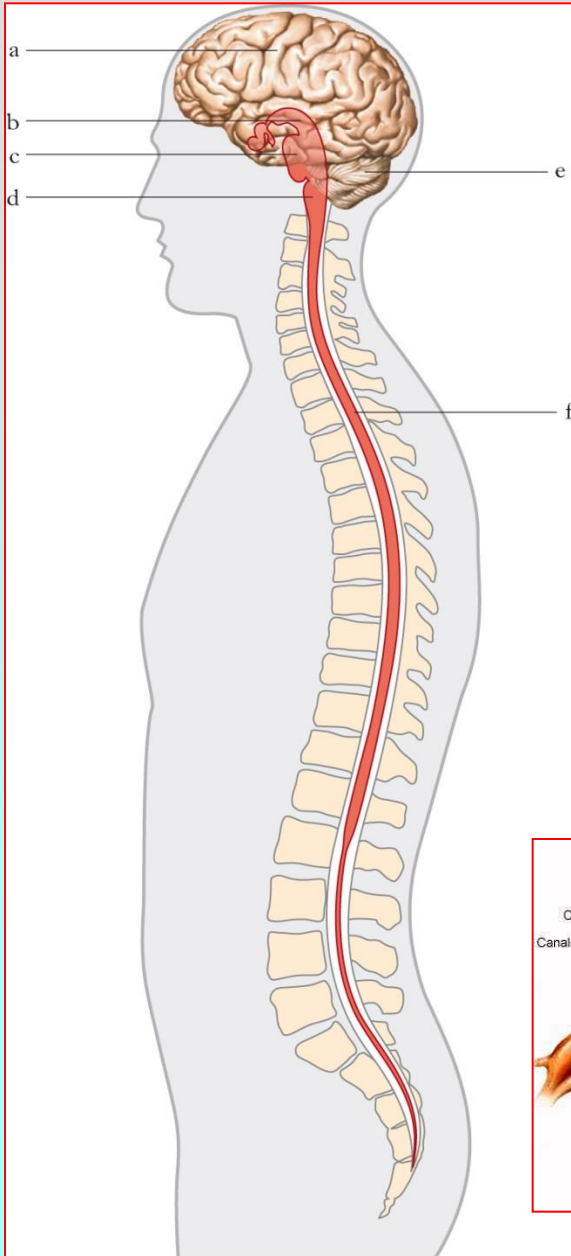
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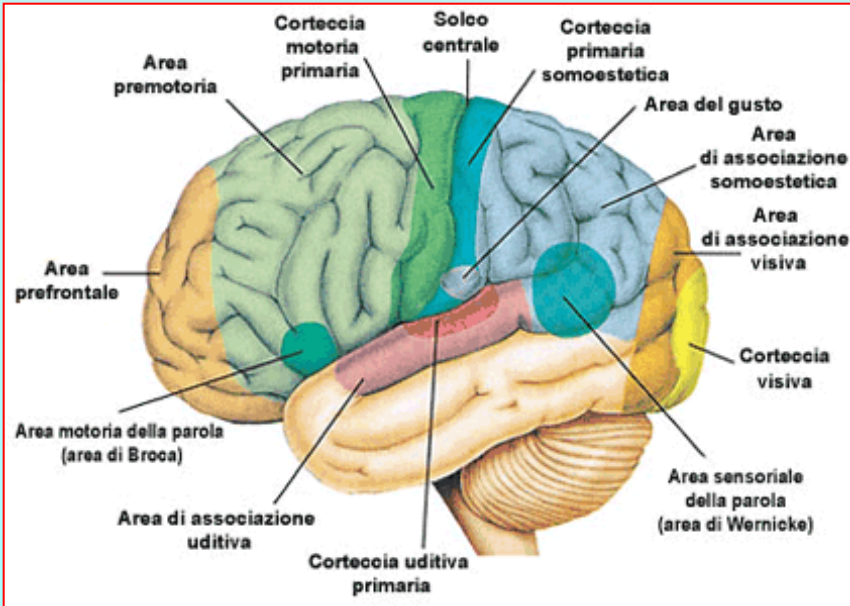
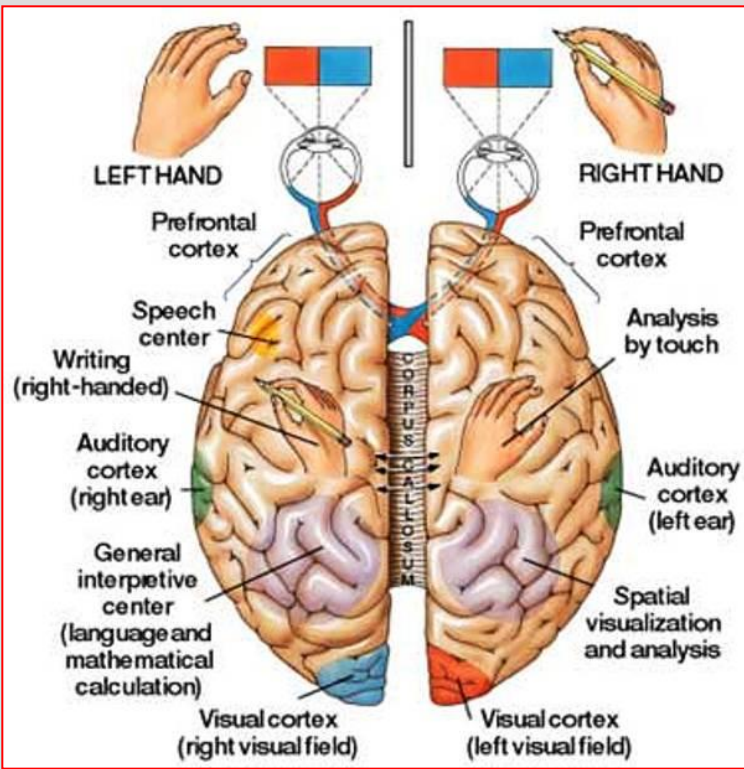


# Sistema Nervoso Centrale

# anatomia del SNC



# inquadramento e peculiarità



**Neuroni**

- Sensibilità/motricità
- Immagazzinamento
- Integrazione/controllo

**Astrociti**

- Metabolismo
- Detossificazione
- Funzione di barriera

**Oligodendrociti**

- Rivestimento mielinico

**Cell. endodiali**

- Riv. ventricoli, canale midollare
- Assorb./produz. liquor

**Microciti**

- Fagocitosi

## neoplasie astrocitarie

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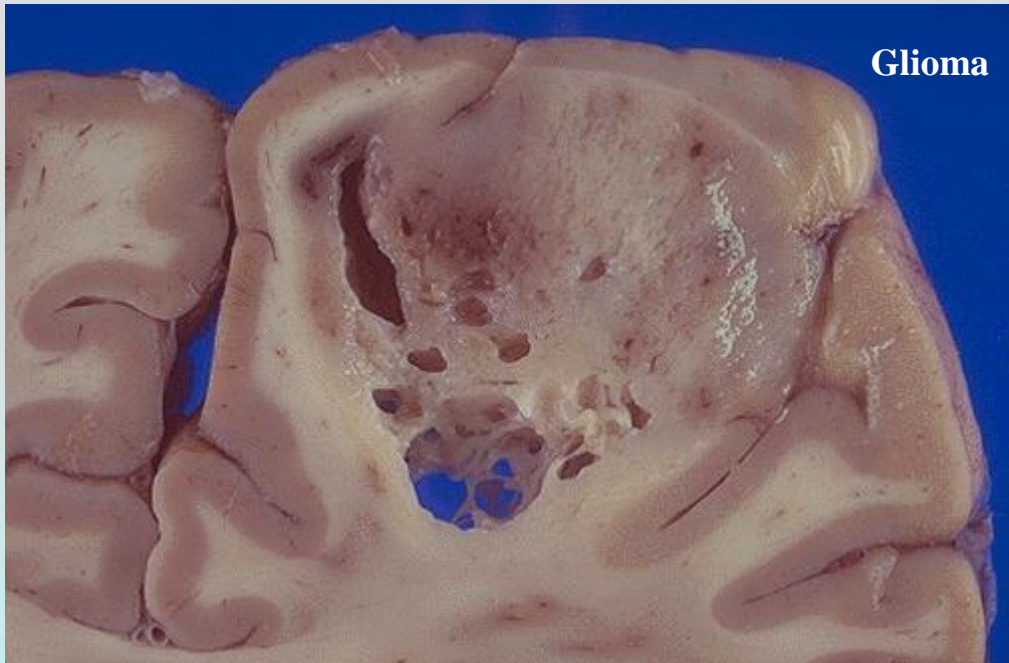
## neoplasie del tessuto neuroepiteliale



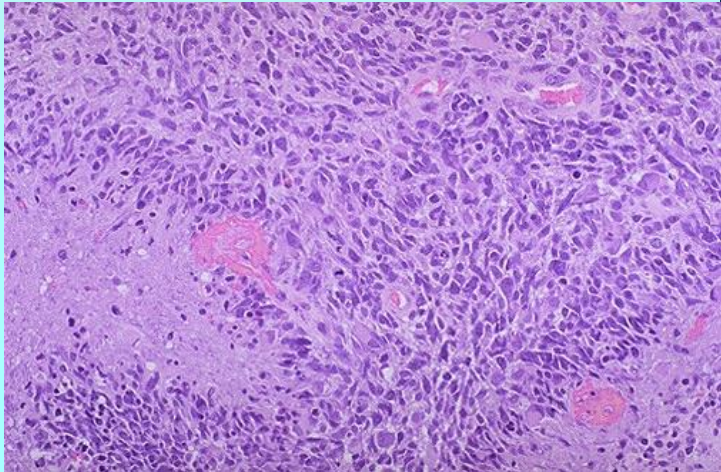
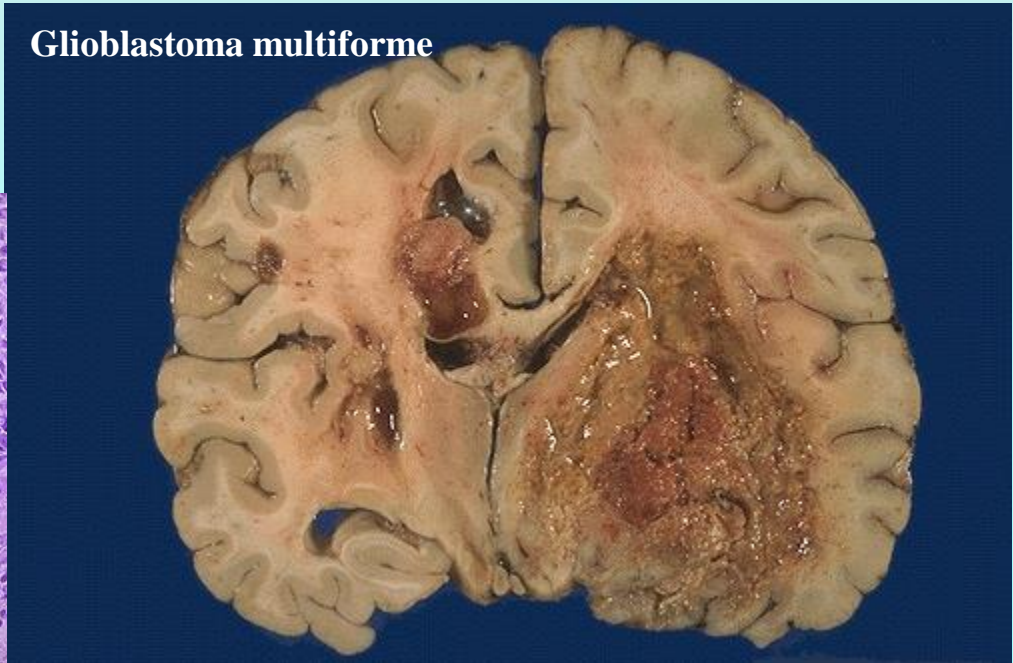
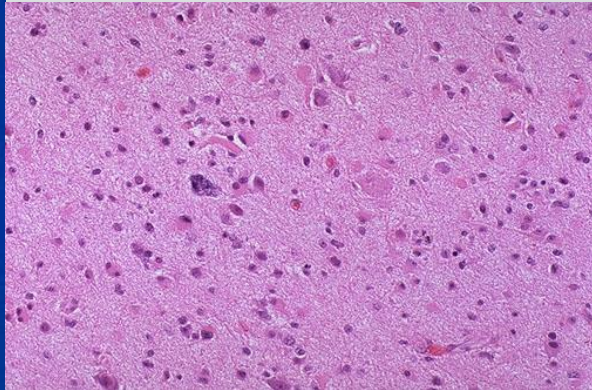
- |                                |        |
|--------------------------------|--------|
| • Astrocitoma pilocitico       | 9421/1 |
| • A. pilomixioide              | 9425/3 |
| • A. subepend. a cell. giganti | 9384/1 |
| • Xantoastrocitoma pleomorfo   | 9424/3 |
| • Astrocitoma diffuso          | 9400/3 |
| • A. fibrillare                | 9420/3 |
| • A. gemistocitico             | 9411/3 |
| • A. protoplasmatico           | 9410/0 |
| • Astrocitoma anaplastico      | 9401/3 |
| • Glioblastoma                 | 9440/0 |
| • GB a cell. giganti           | 9441/3 |
| • Gliosarcoma                  | 9442/0 |
| • Gliomatosis cerebri          | 9381/3 |

- ### Caratteri clinico-biologici (gliomi)
- Principale gruppo di neoplasie del SNC
  - Comprende astrocitomi, oligodendrogliomi ed ependimomi
- ### Caratteri clinico-biologici (astrocitomi)
- 80% dei t. primitivi del SNC negli adulti
  - Forma «pilocitica» giovanile
  - Localizzazione principalmente emisferica (anche mediale, cerebellare, spinale)
  - Più frequente in IV-VI decade
  - Segni e sintomi caratteristici (cefalea, segni neurologici)
  - Ampio spettro di differenziazione/prognosi
  - Progressione basso >>> alto grado

	I	II	III	IV
<b>Astrocytic tumours</b>				
Subependymal giant cell astrocytoma	•			
Pilocytic astrocytoma	•			
Pilomyxoid astrocytoma		•		
Diffuse astrocytoma		•		
Pleomorphic xanthoastrocytoma		•		
Anaplastic astrocytoma			•	
Glioblastoma				•
Giant cell glioblastoma				•
Gliosarcoma				•



gliomi



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**neoplasie del tessuto neuroepiteliale**



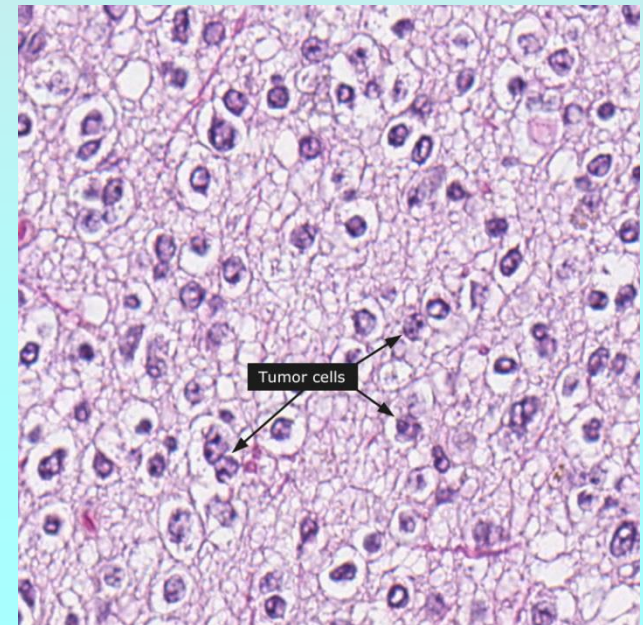
**neoplasie oligodendrogliali**

- Oligodendroglioma 9450/3
- Oligodendr. anaplastico 9451/3

**neoplasie oligoastrocitarie**

- Oligoastrocitoma 9382/3
- Oligoastroc. anaplastico 9382/3

	I	II	III	IV
<b>Oligodendroglial tumours</b>				
Oligodendroglioma		•		
Anaplastic oligodendroglioma			•	
<b>Oligoastrocytic tumours</b>				
Oligoastrocytoma		•		
Anaplastic oligoastrocytoma			•	



## neoplasie dei plessi corioidei

- Papilloma dei pl. corioidei **9390/0**
- Papill. dei p.c. atipico **9390/1**
- Carcinoma dei pl. corioidei **9390/3**

Choroid plexus tumours	I	II	III	IV
Choroid plexus papilloma	•			
Atypical choroid plexus papilloma		•		
Choroid plexus carcinoma			•	

## neoplasie del tessuto neuroepiteliale

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Ependymal tumours	I	II	III	IV
Subependymoma	•			
Myxopapillary ependymoma	•			
Ependymoma		•		
Anaplastic ependymoma			•	

## neoplasie ependimali

## altre neopl. neuroepiteliali

- Astroblastoma **9430/3**
- Glioma cordoide del III ventr. **9440/1**
- Glioma angiocentrico **9431/1**

Other neuroepithelial tumours	I	II	III	IV
Angiocentric glioma	•			
Chordoid glioma of the third ventricle		•		

- Subependimoma **9383/1**
- Ependimoma mixopapillare **9394/1**
- Ependimoma **9391/3**
  - E. cellulare **9391/3**
  - E. papillare **9393/0**
  - E a cellule chiare **9391/3**
  - E. tanicitico **9391/3**
- Ependimoma anaplastico **9392/3**



## neoplasie neuronali e miste neurono-gliali

- Gangliocitoma cerebell. displastico
- Astrocitoma/ganglioglioma desmoplastico infantile
- Tumore neuroepiteliale disembrionoplastico
- Gangliocitoma
- Ganglioglioma
- Ganglioglioma anaplastico
- Neurocitoma centrale
- Neurocitoma extraventricolare
- Liponeurocitoma cerebellare
- Tumore papillare glioneuronale
- Tumore glioneuronale del IV ventricolo formante rosette
- Paraganglioma

9430/0  
9412/1  
9413/0  
9492/0  
9505/1  
9505/3  
9506/1  
9506/1  
9506/1  
9509/1  
9509/1  
8680/1

## neoplasie del tessuto neuroepiteliale



### Caratteri clinico-biologici

- Costituiti da cellule neuronali «normali»
- Più freq. tra le forme «pure»: ganglioglioma
- Spesso compresenza cell. Gliali
- Crescita lenta
- Progressione rapida per anaplasia gliale
- Manifestazioni neurologiche (epilessia)
- Terapia chirurgica

	I	II	III	IV
<b>Neuronal and mixed neuronal-glial tumours</b>				
Gangliocytoma	•			
Ganglioglioma	•			
Anaplastic ganglioglioma				
Desmoplastic infantile astrocytoma and ganglioglioma	•			
Dysembryoplastic neuroepithelial tumour	•			

	I	II	III	IV
<b>Neuronal and mixed neuronal-glial tumours</b>				
Central neurocytoma		•		
Extraventricular neurocytoma		•		
Cerebellar liponeurocytoma		•		
Paraganglioma of the spinal cord	•			
Papillary glioneuronal tumour	•			
Rosette-forming glioneuronal tumour of the fourth ventricle	•			



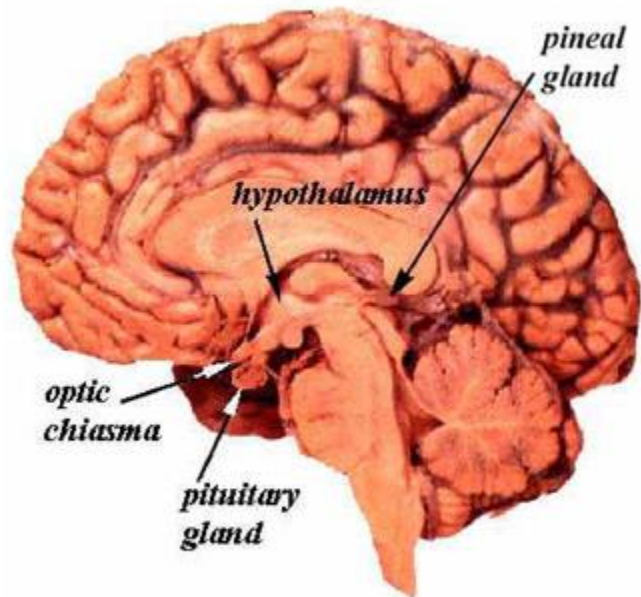
# neoplasie del tessuto neuroepiteliale



## neoplasie della regione pineale

- Pineocitoma 9361/1
- T. pineale parench. a diff. intermedia 9362/3
- Pineoblastoma 9362/3
- Tumore papillare della reg. pineale 9395/3

Pineal tumours	I	II	III	IV
Pineocytoma	•			
Pineal parenchymal tumour of intermediate differentiation		•	•	
Pineoblastoma				•
Papillary tumour of the pineal region		•	•	



## neoplasie embrionali

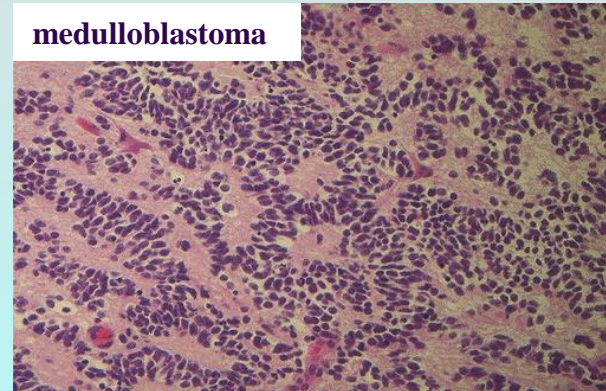
- Medulloblastoma
  - M. desmoplastico/nodulare
  - M. con nodularità estensive
  - M. anaplastico
  - M. a grandi cellule
- Tumore primitivo neuroectodermico del SNC
  - Neuroblastoma del SNC
  - Ganglioneuroblastoma del SNC
  - Medulloepitelioma
  - Ependimoblastoma
- T. atipico teratoide/rabdoide

9470/3  
9471/3  
9471/3  
9474/3  
9474/3  
9473/3  
9500/3  
9490/3  
9501/3  
9392/3  
9508/3

## neoplasie del tessuto neuroepiteliale



medulloblastoma

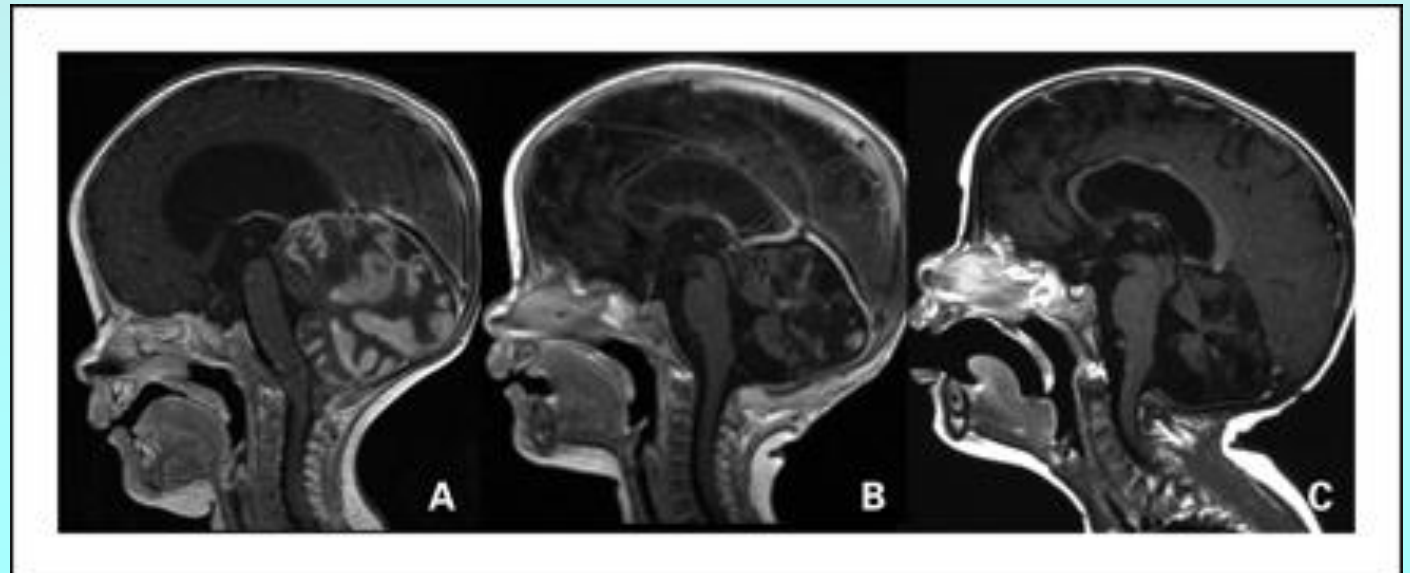


Embryonal tumours	I	II	III	IV
Medulloblastoma				•
CNS primitive neuroectodermal tumour (PNET)				•
Atypical teratoid / rhabdoid tumour				•



### Caratteri clinico-biologici (medulloblastoma)

- Insorgenza nei bambini
- Localizzazione cerebellare (mediale)
- Espressione di markers neuronali e gliali
- Aspetto indifferenziato
- Derivazione «embrionale»
- Comportamento aggressivo
- Altamente radiosensibile
- Prognosi discreta (OS 75% a 5 anni post chirurgia e rx terapia)
- Forme simili (CNS PNET) anche sovratentoriali differenti da MB e da PNET periferici



### The 2007 WHO Classification of Tumours of the Central Nervous System

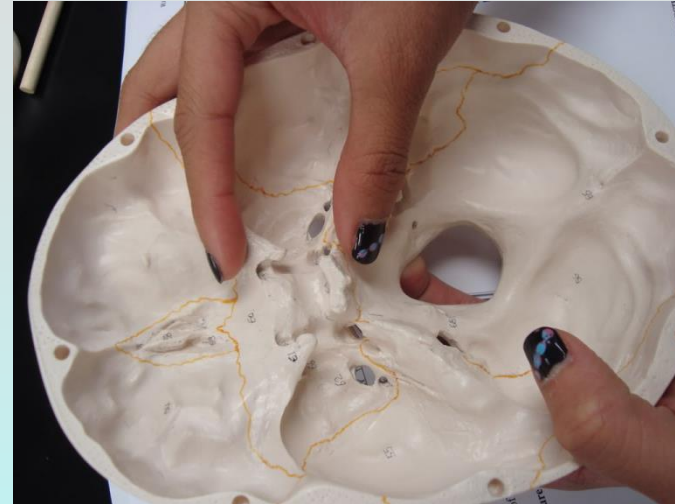
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## neoplasie della sella



### tumori della regione sellare

- **Craniofaringioma** **9350/1**
  - Adamantinomatoso **9351/1**
  - Papillare **9352/1**
- **Tum. a cellule granulari** **9582/0**
- **Pituicitoma** **9432/1**
- **Oncocitoma a cell. fusate dell'adenipofisi** **8290/1**



Tumours of the sellar region	I	II	III	IV
Craniopharyngioma	•			
Granular cell tumour of the neurohypophysis	•			
Pituicytoma	•			
Spindle cell oncocytoma of the adenohypophysis	•			

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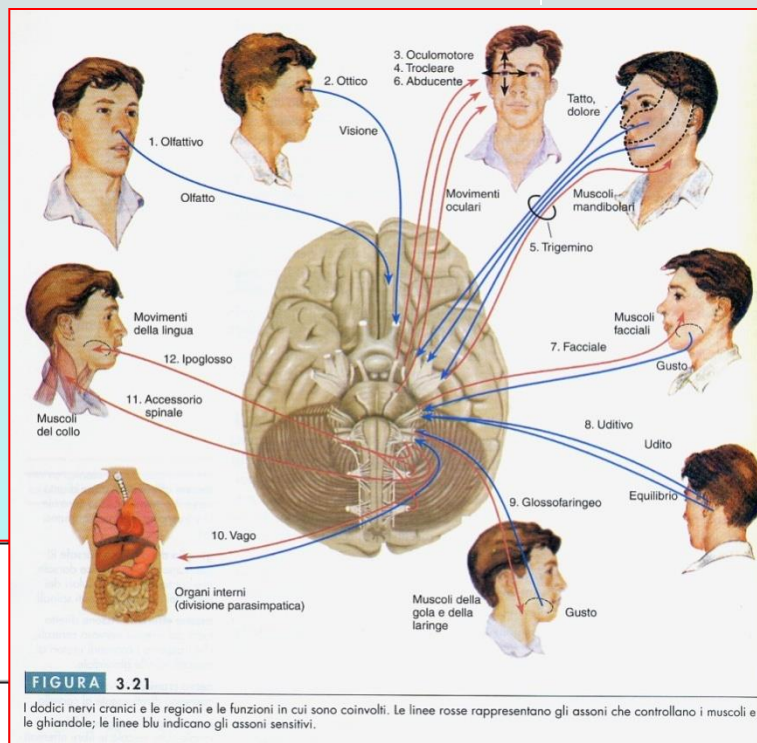
# neoplasie dei nervi



## Neoplasie dei nervi craniali e paraspinali

- Schwannoma
  - S. cellulare
  - S. plessiforme
  - S. melanotico
- Neurofibroma
  - N. plessiforme
- Perineurioma, NAS
  - P. maligno
- Tum. maligno delle guaine nervose periferiche (MPNST)
  - MPNST epitelioido
  - MPNST a differenz. mesenchimale
  - MPNST melanotico
  - MPNST con differenziazione ghiandolare

- 9560/0
- 9560/0
- 9560/0
- 9560/0
- 9540/0
- 9550/0
- 9571/0
- 9571/3
- 9540/3
- 9540/3
- 9540/3
- 9540/3



Tumours of the cranial and paraspinal nerves	I	II	III
Schwannoma	•		
Neurofibroma	•		
Perineurioma	•	•	•
Malignant peripheral nerve sheath tumour (MPNST)		•	•

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Webster K. Cavenee · Peter C. Burger · Anne Jouvett ·  
Bernd W. Scheithauer · Paul Kleihues

## neoplasie del tessuto neuroepiteliale



### nuove entità

• Glioma angiocentrico	9431/1	I
• Papilloma atipico del pl. corioidei	9390/1	II
• Neurocitoma extraventricolare	9506/1	II
• Tumore papillare glioneuronale (PGNT)	9509/1	I
• Tumore glioneuronale del IV ventr. formante rosette	9509/1	I
• Tumore papillare della regione pineale	9395/3	II/III
• Pituitoma	9411/3	I
• Oncocitoma a cell. fusate dell'adenoipofisi	8290/1	II

### nuove entità

• Astrocitoma pilomixoide	9425/3	II
• Medulloblastoma anaplastico	9474/3	IV
• Medulloblastoma con nodularità estensive	9471/3	IV



# altre neoplasie del SNC

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## tumori delle cellule germinali

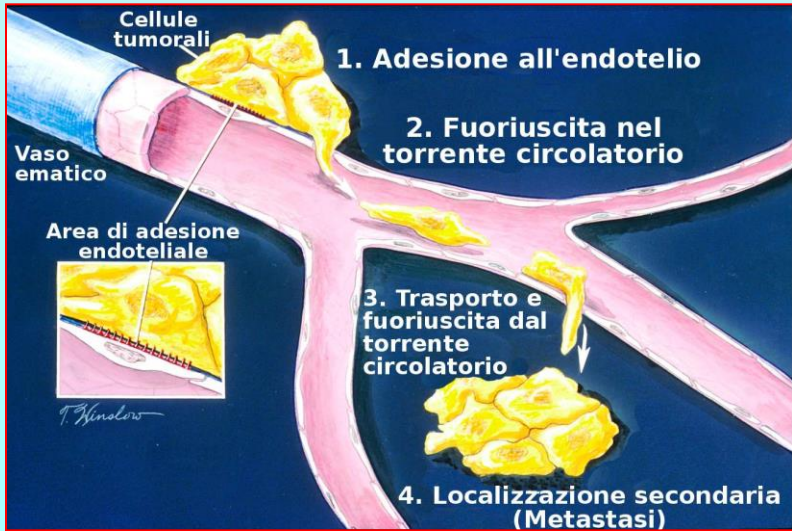
• Germinoma	9640/3
• Carcinoma embrionale	9070/3
• Tumore del sacco vitellino	9071/3
• Coriocarcinoma	9100/3
• Teratoma	9080/1
• Maturo	9080/0
• Immaturo	9080/3
• Con trasf. maligna	9080/3
• Tum. germ. a cell. miste	9085/3

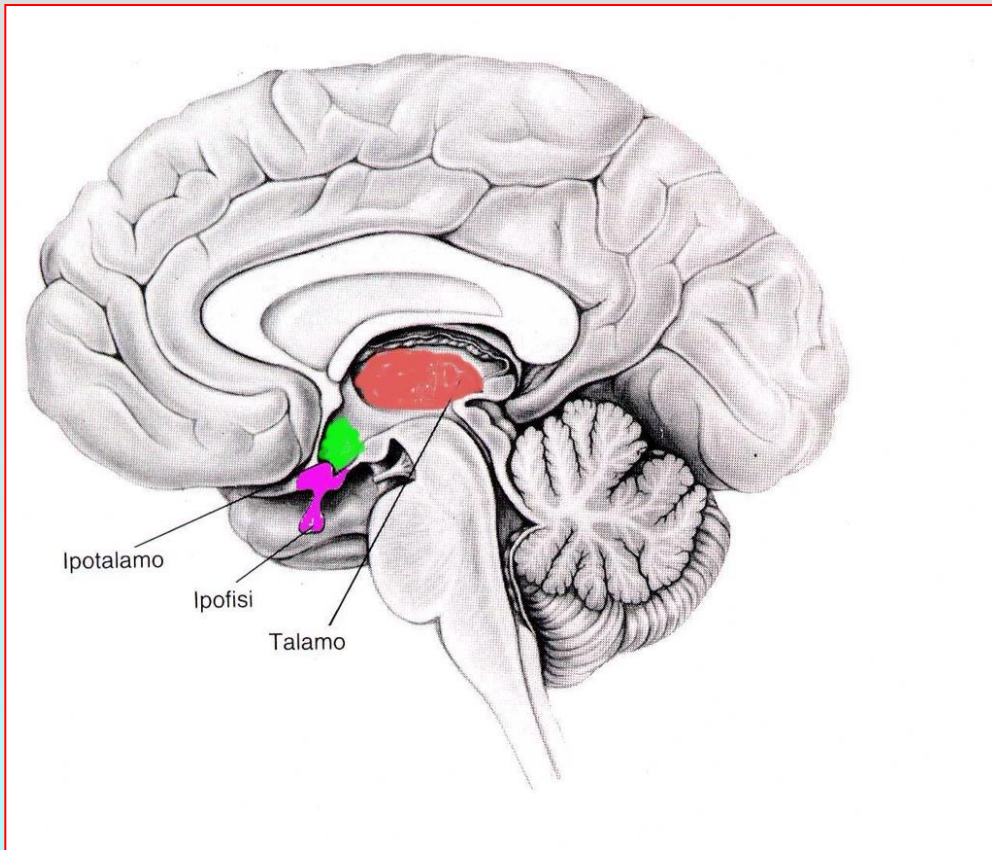
## linfomi e neopl. emopoietiche

• Linfoma maligno	9590/3
• Plasmocitoma	9731/3
• Sarcoma granulocitico	9930/3

## localizzazioni metastatiche

• Carcinomi	80%
• Polmome	
• Mammella	
• Rene	
• Melanoma	
• Gastrointestino	

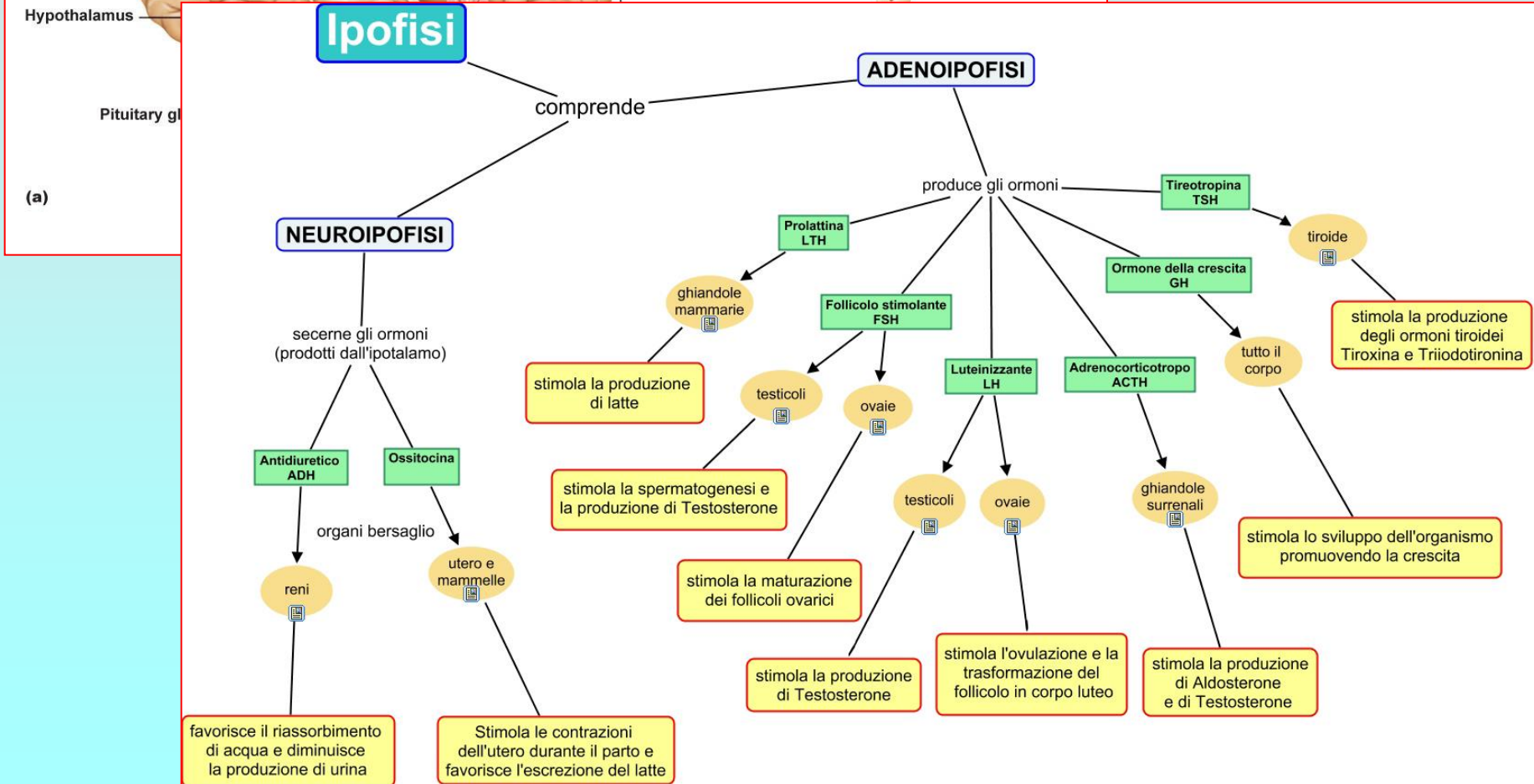
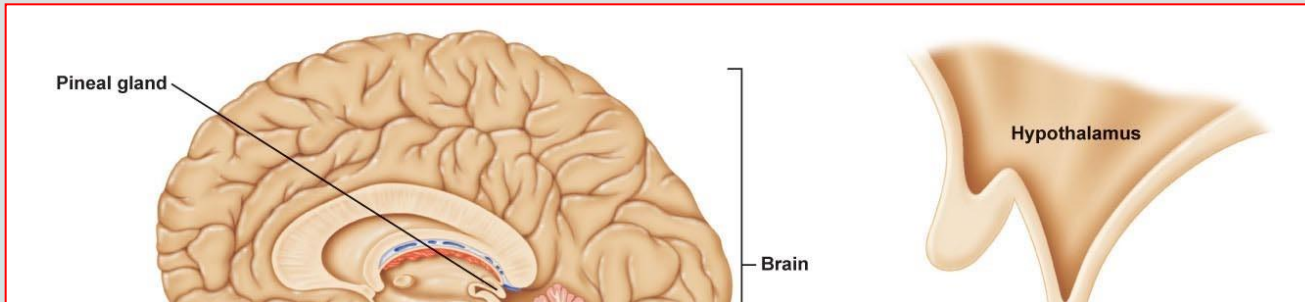




# Ipofisi



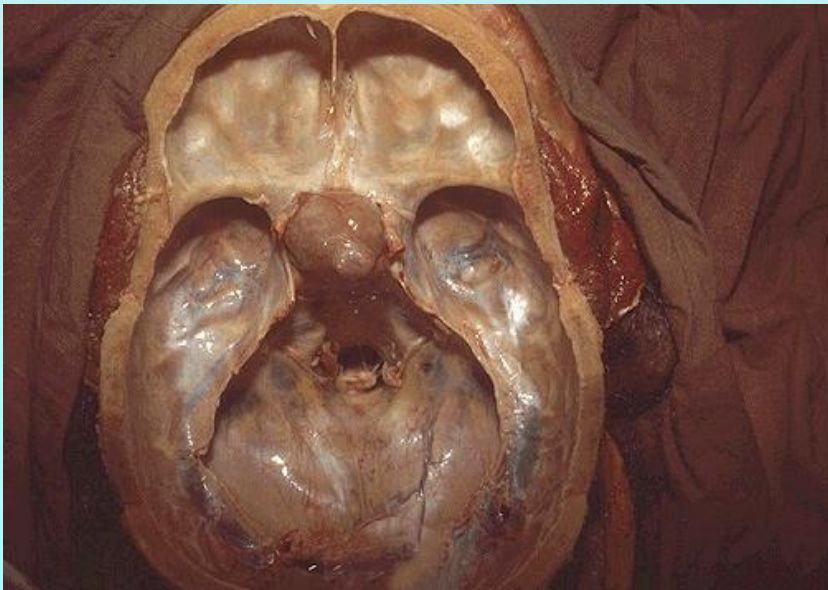
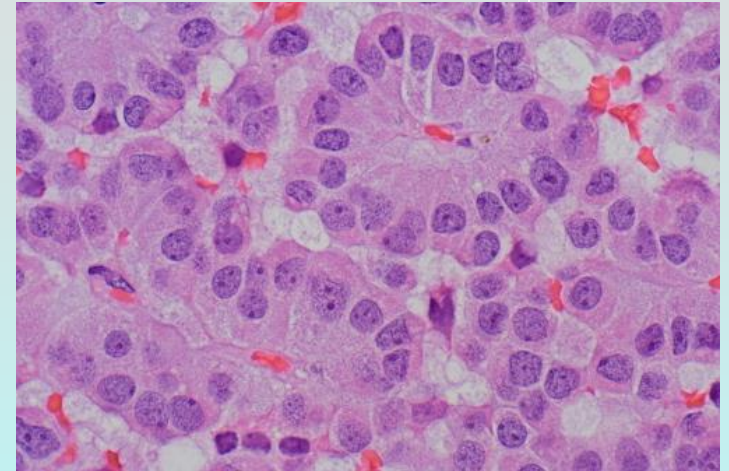
# neoplasie dell'ipofisi





### Tumori ipofisari

- |   |        |
|---|--------|
| • Tum. secernenti prolattina (PRL)                | 8271/0 |
| • Tum. secernenti ormone adenocorticotropo (ACTH) | -      |
| • Tum. secernente ormone della crescita (GH)      | -      |
| • Tum. Secernente tireotropina (TSH)              | -      |
| • Tum. Secernente gonadotropine (FSH-LH)          | -      |
| • Adenomi pluriormonali                           | -      |
| • Adenomi non funzionanti                         | 8272/0 |
| • Tumori oncocitari                               | 8290/0 |
| • Carcinomi                                       | 8272/3 |
| • Altri tumori                                    |        |
| • Tumori metastatici                              |        |



### Clinica

- Produzione inappropriata di ormoni
- Deficit del campo visivo
- Paralisi nervi cranici
- Epilessia temporale
- Idrocefalo
- Rinorrea (liquorrea)



**Grazie!**