

Клиническая диагностика новообразований ЦНС



СКОГАРЕВА НАТАЛЬЯ

ЛФ, 4 КУРС, ГР. 1533

ТОМСК, 2018

WHO classification of tumours of the central nervous system

Diffuse astrocytic and oligodendroglial tumours

Diffuse astrocytoma, IDH-mutant 9400/3
 Gemistocytic astrocytoma, IDH-mutant 9411/3
Diffuse astrocytoma, IDH-wildtype 9400/3
 Diffuse astrocytoma, NOS 9400/3

Anaplastic astrocytoma, IDH-mutant 9401/3
Anaplastic astrocytoma, IDH-wildtype 9401/3
 Anaplastic astrocytoma, NOS 9401/3

Glioblastoma, IDH-wildtype 9440/3
 Giant cell glioblastoma 9441/3
 Gliosarcoma 9442/3
Epithelioid glioblastoma 9440/3
 Glioblastoma, IDH-mutant 9445/3*
 Glioblastoma, NOS 9440/3

Diffuse midline glioma, H3 K27M-mutant 9385/3*

Oligodendroglioma, IDH-mutant and 1p/19q-codeleted 9450/3
 Oligodendroglioma, NOS 9450/3

Anaplastic oligodendroglioma, IDH-mutant and 1p/19q-codeleted 9451/3
Anaplastic oligodendroglioma, NOS 9451/3

Oligoastrocytoma, NOS 9382/3
Anaplastic oligoastrocytoma, NOS 9382/3

Other astrocytic tumours

Pilocytic astrocytoma 9421/1
 Piloxyoid astrocytoma 9425/3
 Subependymal giant cell astrocytoma 9384/1
 Pleomorphic xanthoastrocytoma 9424/3
 Anaplastic pleomorphic xanthoastrocytoma 9424/3

Ependymal tumours

Subependymoma 9383/1
 Myxopapillary ependymoma 9394/1
 Ependymoma 9391/3
 Papillary ependymoma 9393/3
 Clear cell ependymoma 9391/3
 Tancytic ependymoma 9391/3
 Ependymoma, *RELA* fusion-positive 9396/3*
 Anaplastic ependymoma 9392/3

Other gliomas

Chordoid glioma of the third ventricle 9444/1
 Angiocentric glioma 9431/1
 Astroblastoma 9430/3

Choroid plexus tumours

Choroid plexus papilloma 9390/0
 Atypical choroid plexus papilloma 9390/1
 Choroid plexus carcinoma 9390/3

Neuronal and mixed neuronal-glial tumours

Dysembryoplastic neuroepithelial tumour 9413/0
 Gangliocytoma 9492/0
 Ganglioglioma 9505/1
 Anaplastic ganglioglioma 9505/3
 Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease) 9493/0
 Desmoplastic infantile astrocytoma and ganglioglioma 9412/1
 Papillary glioneuronal tumour 9509/1
 Rosette-forming glioneuronal tumour 9509/1
Diffuse leptomeningeal glioneuronal tumour
 Central neurocytoma 9506/1
 Extraventricular neurocytoma 9506/1
 Cerebellar liponeurocytoma 9506/1
 Paranglioma 8693/1

Tumours of the pineal region

Pineocytoma 9361/1
 Pineal parenchymal tumour of intermediate differentiation 9362/3
 Pineoblastoma 9362/3
 Papillary tumour of the pineal region 9395/3

Embryonal tumours

Medulloblastomas, genetically defined
 Medulloblastoma, WNT-activated 9475/3*
 Medulloblastoma, SHH-activated and *TP53*-mutant 9476/3*
 Medulloblastoma, SHH-activated and *TP53*-wildtype 9471/3
 Medulloblastoma, non-WNT/non-SHH
 Medulloblastoma, group 3 9477/3*
 Medulloblastoma, group 4
 Medulloblastomas, histologically defined
 Medulloblastoma, classic 9470/3
 Medulloblastoma, desmoplastic/nodular 9471/3
 Medulloblastoma with extensive nodularity 9471/3
 Medulloblastoma, large cell / anaplastic 9474/3
 Medulloblastoma, NOS 9470/3
 Embryonal tumour with multilayered rosettes, *C19MC*-altered 9478/3*
Embryonal tumour with multilayered rosettes, NOS 9478/3
 Medulloepithelioma 9501/3
 CNS neuroblastoma 9500/3
 CNS ganglioneuroblastoma 9490/3
 CNS embryonal tumour, NOS 9473/3
 Atypical teratoid/rhabdoid tumour 9508/3
CNS embryonal tumour with rhabdoid features 9508/3

Tumours of the cranial and paraspinal nerves

Schwannoma 9560/0
 Cellular schwannoma 9560/0
 Plexiform schwannoma 9560/0

Melanotic schwannoma	9560/1	Osteochondroma	9210/0
Neurofibroma	9540/0	Osteosarcoma	9180/3
Atypical neurofibroma	9540/0		
Plexiform neurofibroma	9550/0	Melanocytic tumours	
Perineurioma	9571/0	Meningeal melanocytosis	8728/0
Hybrid nerve sheath tumours		Meningeal melanocytoma	8728/1
Malignant peripheral nerve sheath tumour	9540/3	Meningeal melanoma	8720/3
Epithelioid MPNST	9540/3	Meningeal melanomatosis	8728/3
MPNST with perineurial differentiation	9540/3		
		Lymphomas	
Meningiomas		Diffuse large B-cell lymphoma of the CNS	9680/3
Meningioma	9530/0	Immunodeficiency-associated CNS lymphomas	
Meningothelial meningioma	9531/0	AIDS-related diffuse large B-cell lymphoma	
Fibrous meningioma	9532/0	EBV-positive diffuse large B-cell lymphoma, NOS	
Transitional meningioma	9537/0	Lymphomatoid granulomatosis	9766/1
Psammomatous meningioma	9533/0	Intravascular large B-cell lymphoma	9712/3
Angiomatous meningioma	9534/0	Low-grade B-cell lymphomas of the CNS	
Microcystic meningioma	9530/0	T-cell and NK/T-cell lymphomas of the CNS	
Secretory meningioma	9530/0	Anaplastic large cell lymphoma, ALK-positive	9714/3
Lymphoplasmacyte-rich meningioma	9530/0	Anaplastic large cell lymphoma, ALK-negative	9702/3
Metaplastic meningioma	9530/0	MALT lymphoma of the dura	9699/3
Chordoid meningioma	9538/1		
Clear cell meningioma	9538/1	Histiocytic tumours	
Atypical meningioma	9539/1	Langerhans cell histiocytosis	9751/3
Papillary meningioma	9538/3	Erdheim-Chester disease	9750/1
Rhabdoid meningioma	9538/3	Rosai-Dorfman disease	
Anaplastic (malignant) meningioma	9530/3	Juvenile xanthogranuloma	
		Histiocytic sarcoma	9755/3
		Germ cell tumours	
Mesenchymal, non-meningothelial tumours		Germinoma	9064/3
Solitary fibrous tumour / haemangiopericytoma**		Embryonal carcinoma	9070/3
Grade 1	8815/0	Yolk sac tumour	9071/3
Grade 2	8815/1	Choriocarcinoma	9100/3
Grade 3	8815/3	Teratoma	9080/1
Haemangioblastoma	9161/1	Mature teratoma	9080/0
Haemangioma	9120/0	Immature teratoma	9080/3
Epithelioid haemangi endothelioma	9133/3	Teratoma with malignant transformation	9084/3
Angiosarcoma	9120/3	Mixed germ cell tumour	9085/3
Kaposi sarcoma	9140/3		
Ewing sarcoma / PNET	9364/3	Tumours of the sellar region	
Lipoma	8850/0	Craniopharyngioma	9350/1
Angiolipoma	8861/0	Adamantinomatous craniopharyngioma	9351/1
Hibernoma	8880/0	Papillary craniopharyngioma	9352/1
Liposarcoma	8850/3	Granular cell tumour of the sellar region	9582/0
Desmoid-type fibromatosis	8821/1	Pituicytoma	9432/1
Myofibroblastoma	8825/0	Spindle cell oncocytoma	8290/0
Inflammatory myofibroblastic tumour	8825/1		
Benign fibrous histiocytoma	8830/0		
Fibrosarcoma	8810/3		
Undifferentiated pleomorphic sarcoma / malignant fibrous histiocytoma	8802/3	Metastatic tumours	
Leiomyoma	8890/0		
Leiomyosarcoma	8890/3		
Rhabdomyoma	8900/0		
Rhabdomyosarcoma	8900/3		
Chondroma	9220/0		
Chondrosarcoma	9220/3		
Osteoma	9180/0		

The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) (742A). Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours. The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions. *These new codes were approved by the IARC/WHO Committee for ICD-O. †: Provisional tumour entities. **Grading according to the 2013 WHO Classification of Tumours of Soft Tissue and Bone.

Молекулярные маркеры



Ген IDH

Ко-делеция 1p / 19q

TP53

TERT

Основное звено патогенеза
опухолей ЦНС – снижение
метиляции, ведущее к
нарушению процессов
репарации ДНК.

Как можно выявить маркеры?



Иммуногистохимия

ПЦР

FISH-метод

Диагностика



Общемозговые симптомы:

Гипертензионно-гидроцефальный синдром;
Дислокационный синдром.

Очаговые симптомы:

Эпилептические припадки;
Расстройства чувствительности и движения;
Симптомы поражения ядер ЧМН;
Расстройства высших корковых функций.

Оболочечные симптомы.

Гипертензионно-гидроцефальный синдром

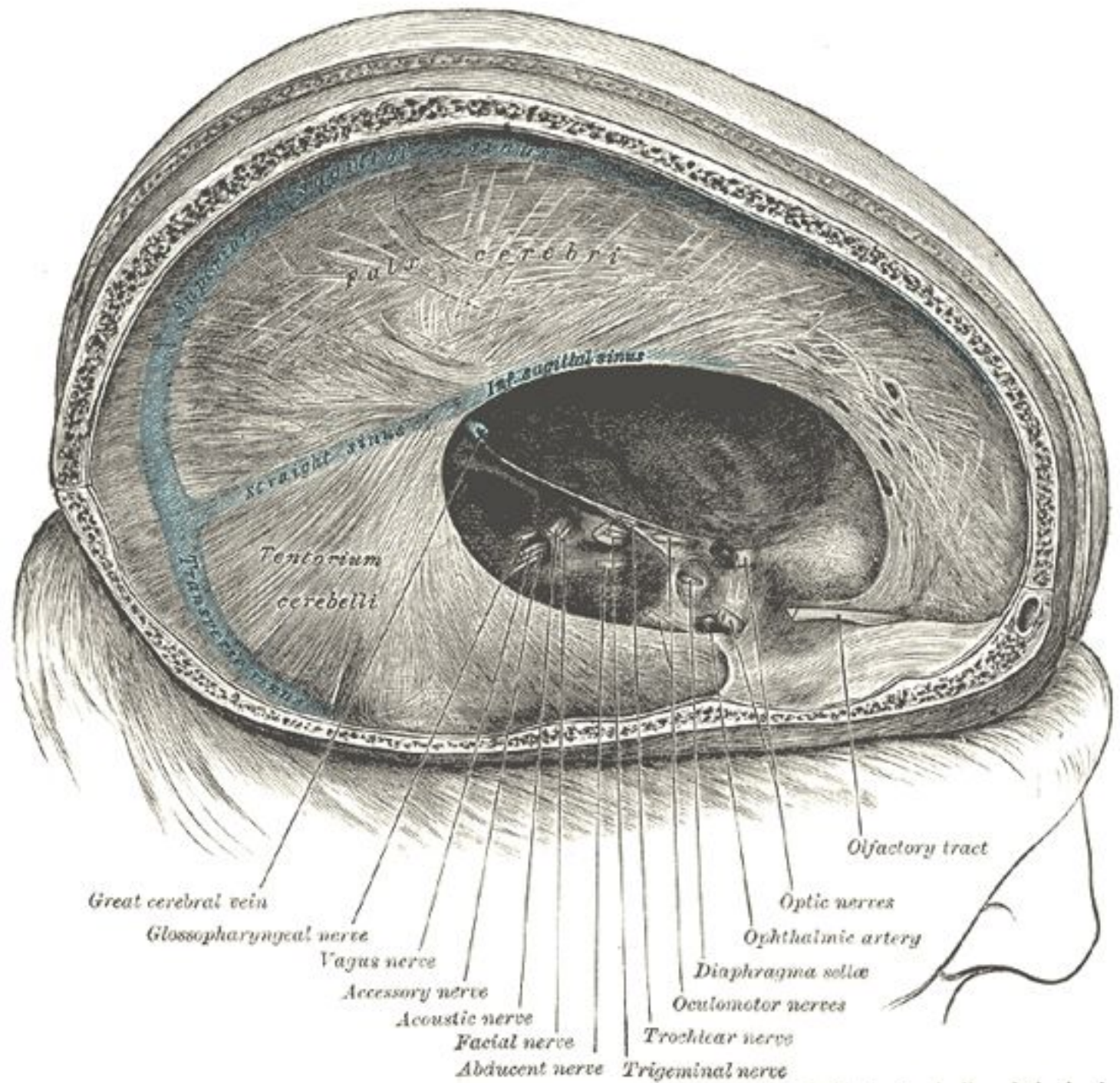


Головная боль

Тошнота

Рвота

Головокружение



Глиобластома

+GD

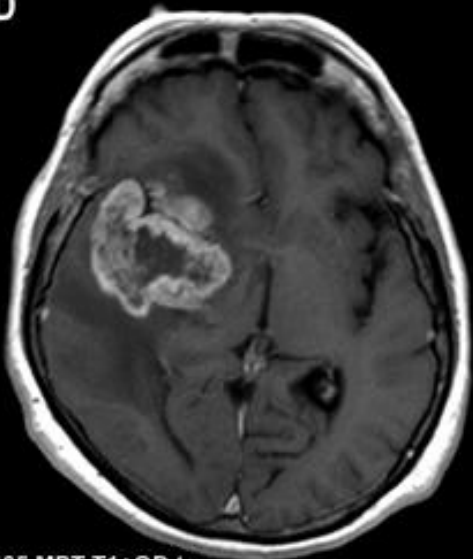


Рис.305 MPT T1+GD tra

+GD

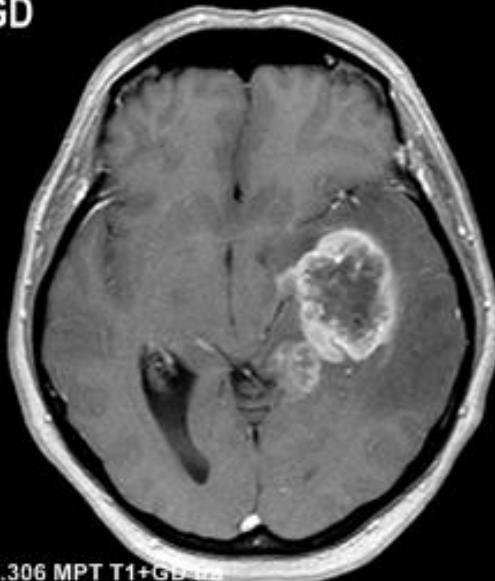


Рис.306 MPT T1+GD tra

+GD



Рис.307 MPT T1+GD tra

+GD

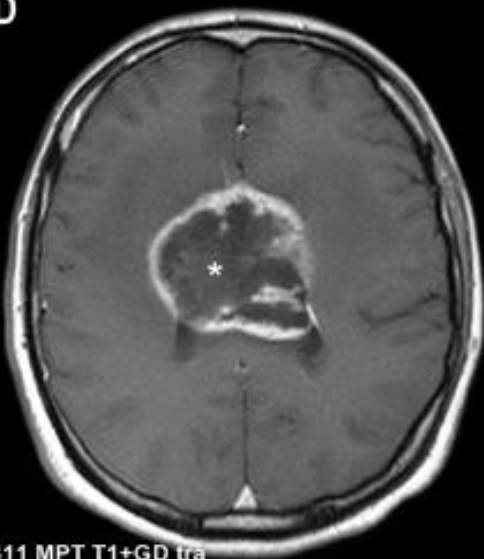


Рис.311 MPT T1+GD tra

+GD



Рис.312 MPT T1+GD cor

+GD

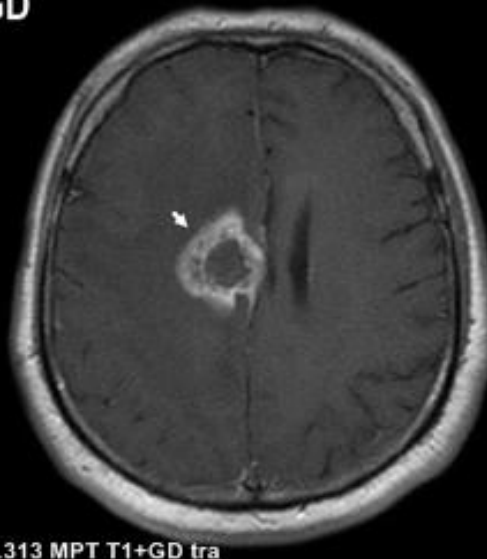
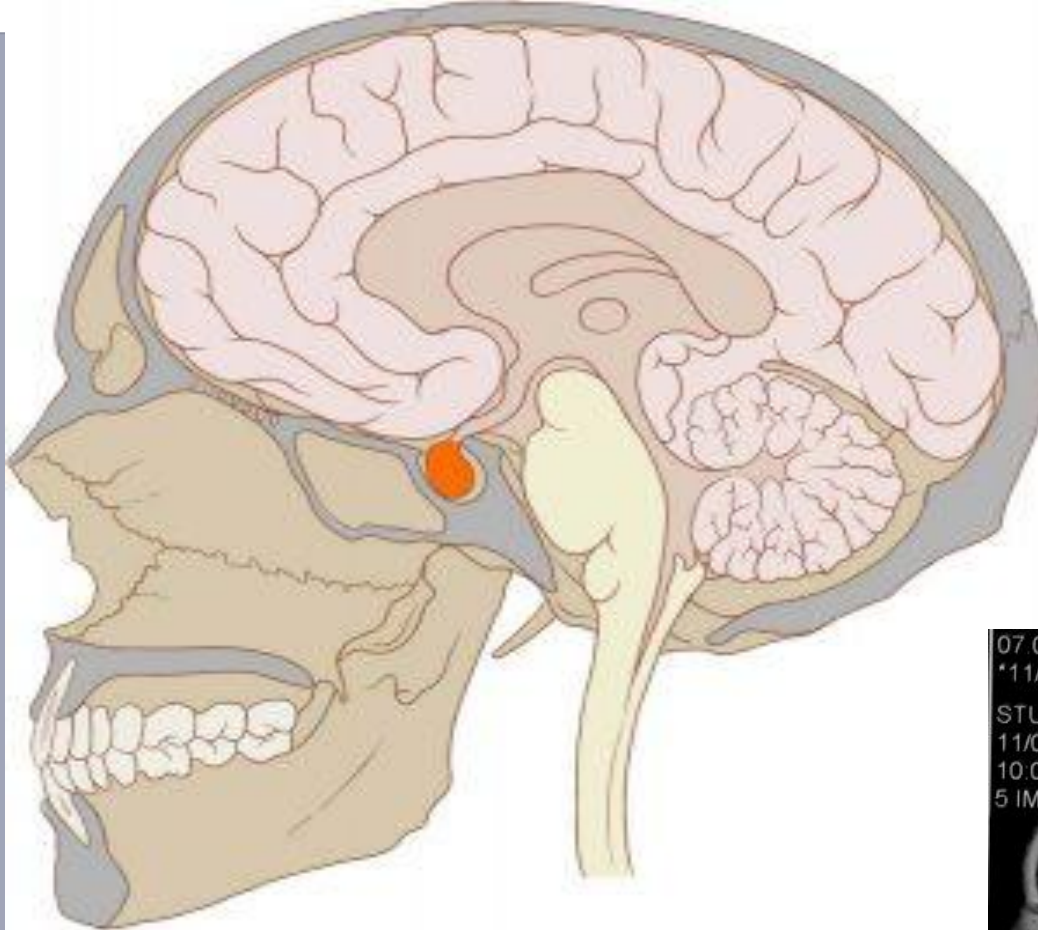
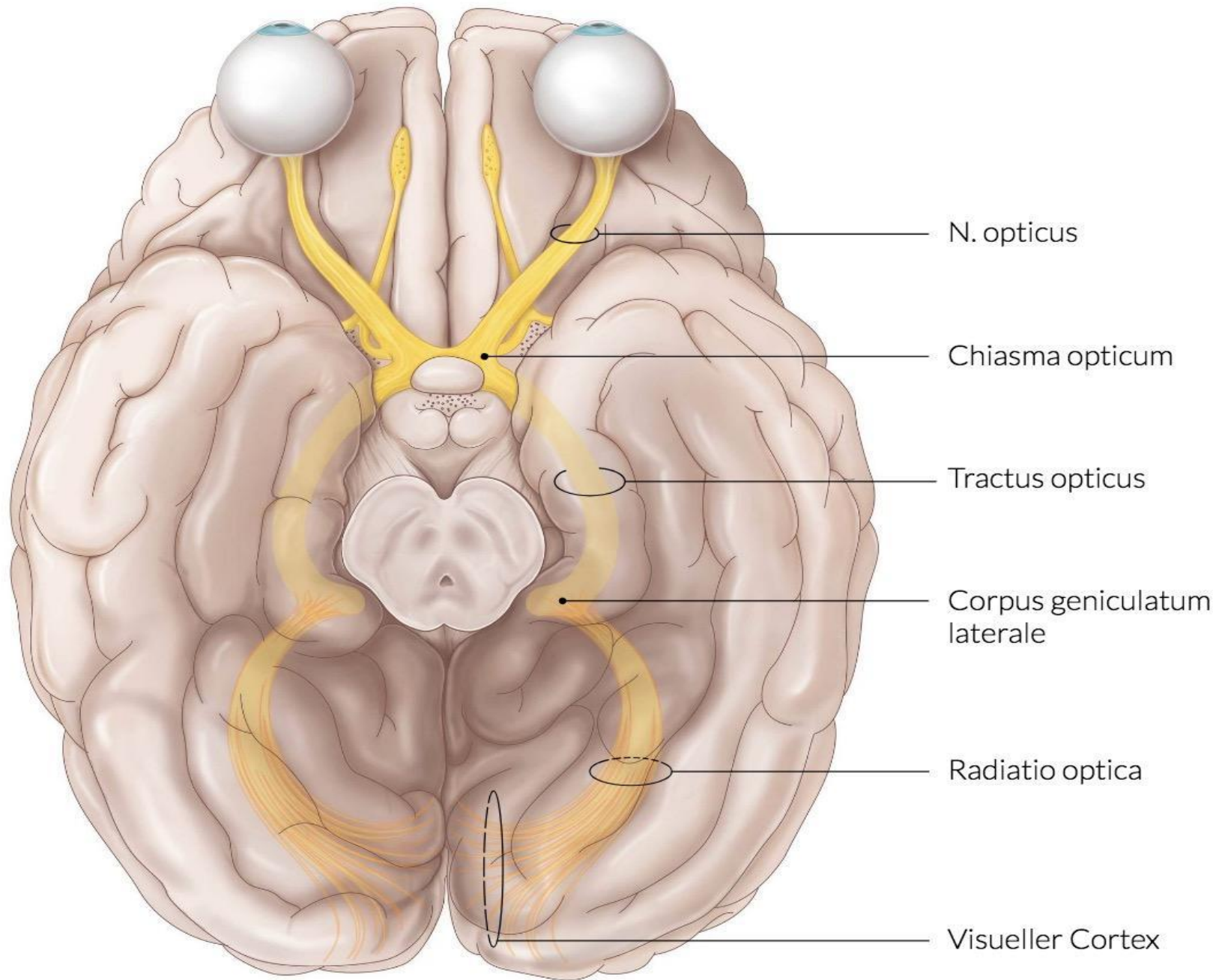


Рис.313 MPT T1+GD tra





Резюме

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