# Epidemiology of brain and spine tumors

Dr Mathangi J 38<sup>th</sup> AROI – ICRO SUN PG Teaching course

## Brain Tumors

- The brain tumors primary and secondary (metastases)
- Metastases more common 10:1
- MC primaries Lung, Breast, Kidney, Colorectal, Melanoma
- Primary BT rise in incidence
- i. better diagnostic ability, better
- ii. access to medical care, and better
- iii. care for the elderly

Continuum (Minneap Minn) 2015;21(2):301-

### Global Incidence

- Malignant brain tumors 4.25 cases per 100,000 person
- Regional variation 6.76 in Europe to 2.81 in Africa
- 6.29 cases per 100,000 in high income countries (HICs), to 4.81 in low and middle-income countries (LMICs).
- Malignant spinal tumors 0.098 cases per 1,00,000 py and varied similarly by region and income group.

**Global incidence of brain and spinal tumors by geographic region and income level based on cancer registry data** J Clin Neurosci. 2019 Aug;66:121-127. doi: 10.1016/j.jocn.2019.05.003. Epub 2019 May 24. Joseph S.Bell<sup>a</sup>

# CBTRUS (+CDC& NCI)

- Brain tumors 2% of all cancers
- AAAIR 23.79 (2012 -2016) 26,070 new cases, 16947 deaths
- Malignant : non malignant 7.08:16.71
- F:M 1: 1.3MC BT : Males: GBM ; Females: Meningiomas
- Hispanics vs Non Hispanics: 21.48 : 24.23
- Meningioma, Pituitary tumors, Craniopharyngioma more in blacks

- Astrocytoma and ODG, Ependymal tumors, Embryonal tumors, Lymphoma, and GCT more in whites

## Variance in Incidence

- Age: brain tumors increases with age
- Younger: Pilocytic astrocytoma, choroid plexus tumors, neuronal tumors, pineal region tumors, and germ cell tumors
- >65 : GBM, Meningiomas
- MBT: 5Y Survival: 33% overall (Anaplastic: 30% and GBM:3%)
- young age, high performance status, and lower pathologic grade
- duration of symptoms, presence of cognitive alterations at diagnosis, posterior fossa location of tumor, and extent of surgical resection.

## ACS statistics 2021

## Brain and other nervous system

#### AT A GLANCE

Estimated new cases, Esti 2021 24,530	mated deaths, 2021 <b>18,600</b>	Incidence rates, 2013- 2017 6.5 Average annual rate per 100,000, age adjusted to the 2000 US standard population.	Death rates, 2014-2018 4.4 Average annual rate per 100,000, age adjusted to the 2000 US standard population. Rates for PR are for 2012-2016.
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### Distribution of All Primary Brain and Other CNS Tumors



\* All or some of this histology is included in the CBTRUS definition of gliomas, including ICD-O-3 histology codes 9380-9384 and, 9391-9460 (Table 2). a. Percentages may not add up to 100% due to rounding. b. Includes oligodendroglioma and anaplastic oligodendroglioma (Table 2). c. Includes pilocytic astrocytoma, diffuse astrocytoma, anaplastic astrocytoma, and unique astrocytoma variants (Table 3). d. Includes glioma malignant, NOS, choroid plexus tumors, other neuroepithelial tumors, neuronal and mixed neuronal-glial tumors, tumors of the pineal region, other tumors of cranial and spinal nerves, mesenchymal tumors, primary melanocytic lesions, other neoplasms related to the meninges, other hematopoietic neoplasms, hemangioma, neoplasm, unspecified, and all other (Table 2).

Neuro Oncol, Volume 21, Issue Supplement\_5, October 2019, Pages v1-v100, https://doi.org/10.1093/neuonc/noz150

## Incidence – India - Globocon

- Brain tumors expected statistics for year 2021
- New cases: 31,460 new cases
- 26,656 deaths
- 5 year prevalence: for all ages: 5.39 per 100,000

J Neurooncol (2008) 87:111–114 DOI 10.1007/s11060-007-9487-z

LETTER TO THE EDITOR

## Western vs Indian data

Prospective analysis of incidence of central nervous tumors presenting in a tertiary cancer hospital from India

Rakesh Jalali • Debnarayan Datta

- Younger age at presentation Benign:Malignant:Metastatic 34,37,49.5 years vs 29, 63, 61 years
- GBM at earlier age ( median age 50 years)
- CNS lymphoma one decade earlier different IHC features
- Pilocytic astrocytoma slightly different molecular genetics
- Lower LE in developing world
- lower geriatric age tumors > 65 years (9% vs 35%)
- lower HGG (16% vs 23%)
- higher pediatric tumors(35% vs 17%)

#### **Original Research Article**

Descriptive epidemiology of central nervous system tumors in rural hospital of central India : 5-year experience

Prasheelkumar Premnarayan Gupta<sup>1,\*</sup>, Richa Premnarayan Goyal<sup>2</sup>

<sup>1</sup>Dept. of Neurosurgery, Mahatma Gandhi Institute of Medical Sciences, Sevagram, Maharashtra, India
<sup>2</sup>Dept. of Surgery, Jawaharlal Nehru Medical College, Wardha, Maharashtra, India

- Lower intracranial tumor incidence
- MC were astrocytomas GBMs, second MC were meningiomas
- Sellar region tumors slightly higher
- Lesser percentage of secondaries
- MC spinal tumors were meningiomas and schwannomas



## Pediatric brain tumors

- Most common solid tumors; second common after leukemias
- Slight male preponderance: 1.5:1 (cultural factors, sex hormones)
- Commonly infratentorial
- Lowgrade astrocytomas MC; Medulloblastoma MC Malignant BT
- Others being BSG and ependymoma
- In the supratentorial region:
- Suprasellar or parasellar region: CP, optic/chiasmal/ hypothalamic gliomas
- Pineal gland tumors and hemispheric gliomas

# Challenges with PBT

- Late diagnosis/ misdiagnosis
- Damage to developing brain treatment induced
- Neurocognitive, Psychological, Endocrine
- Tumors carry better prognosis than their adult counterparts
- New classification incorporating molecular charateristics eg. MB

### Distribution in Children (Ages 0–14) of Primary Brain and CNS Tumors by Histology (N =16,044), CBTRUS, 2007–2011



### Figure 3

- Oligoastrocytic Tumors
- Lymphoma
- Pilocytic Astrocytoma
- Embryonal Tumors
- Malignant Glioma
- All "Other" Tumors
- All "Other" Asrocytoma
- Neuronal/Mixed Neuronal-Glial
- Ependymal Tumors
- Nerve Sheath Tumors
- Craniopharyngioma
- Pituitary Tumors
- Germ Cell Tumors
- Glioblastoma
- Meningioma
- Oligodendrogliomas





Indian J Med Paediatr Oncol. 2017 Jan-Mar; 38(1): 10–14. doi: <u>10.4103/0971-5851.203514</u> PMCID: PMC5398098 PMID: <u>28469330</u>

### Profile of Primary Pediatric Brain and Spinal Cord Tumors from North India

Nadia Shirazi, Meenu Gupta,<sup>1</sup> Nowneet Kumar Bhat,<sup>2</sup> Braham Prakash Kalra,<sup>2</sup> Ranjit Kumar,<sup>3</sup> and Manju Saini<sup>4</sup>

Histopathological diagnosis	Number of cases (%)	Mean age (years)	Male:female
Ewing's sarcoma/primitive neuroectodermal tumor	4 (30.76)	12.2	1:3
Ependymoma	3 (23.07)	8.4	2:1
Glioblastoma multiforme	1 (7.69)	12	Female
Non-Hodgkin's lymphoma	2 (15.38)	7.4	1:1
Malignant peripheral nerve sheath tumor	1 (7.69)	17	Male
Schwannoma	1 (7.69)	15.7	Male
—Pilocytic astrocytoma	1 (7.69)	12	Female

### **Original Article**

### Spectrum of pediatric brain tumors in India: A multi-institutional study

Ayushi Jain, Mehar C. Sharma, Vaishali Suri, Shashank S. Kale<sup>1</sup>, A. K. Mahapatra<sup>1</sup>, Medha Tatke<sup>2</sup>, Geeta Chacko<sup>3</sup>, Ashish Pathak<sup>4</sup>, Vani Santosh<sup>5</sup>, Preeta Nair<sup>6</sup>, Nuzhat Husain<sup>7</sup>, Chitra Sarkar

Departments of Pathology and <sup>1</sup>Neurosurgery, All India Institute of Medical Sciences (AIIMS), New Delhi, <sup>2</sup>Department of

#### Table 2: Percentage breakup of various histological subtypes of pediatric CNS tumors

Tumor	AIIMS	NIMHANS	<b>GB</b> Pant	тмн	CSMMU	CMC Vellore	PGIMER	Average
Astrocytoma	33.7	44.1	22.3	28.6	30.6	46.7	37	34.7
MB and PNETs	16.8	19.7	32	29	27.7	10.3	21.6	22.4
Craniopharyngioma	12.7	7.7	13.5	4.5	13.1	8.5	11.5	10.2
Ependymal	8.5	8.5	12.2	19.1	9.4	4.8	6.3	9.8
Nerve sheath	7	4.3	1.3	2.4	2.2	4.6	NA	3.6
Meningeal	5.6	4.3	0.3	3.4	2.2	3.5	NA	3.2
Neuronal and mixed neuronal glial	4.1	2.8	5.2	2.1	0	NA	NA	2.4
Germ cell tumors	2.2	2.2	3.3	1.7	2.2	NA	NA	2
Choroid plexus tumors	1.5	2.6	1.6	1.7	1.5	NA	3.5	1.8
Pineal tumors	0.7	1.4	1.3	1	3	NA	NA	1.3
Oligodendroglioma	0.7	0.9	2.9	1.4	1.5	0	0	1.1
Lymphoma	1	0.5	0.3	0	0	1.1	NA	0.5

Table 4: Frequency of various types of pediatric CNS tumors reported in different countries (in percentage)

Tumor	Brazil <sup>[1]</sup>	Korea <sup>[5]</sup>	Germany <sup>[6]</sup>	Canada <sup>[7]</sup>	Beijing <sup>[8]</sup>	Sweden <sup>[9]</sup>	Morocco <sup>[10]</sup>	Japan <sup>[11]</sup>	India
									(present study)
Astrocytomas	32.5	27.8	41.7	39.4	30.5	51	37.1	35.7	34.7
Oligodendrogliomas	0.9	2.6	1.1	1.7	6.2	0	1.7	0	1.1
Ependymomas	7.4	8.1	10.4	7	5.6	8	12	4.8	9.8
Choroid plexus tumors	3	2.2	NA	2.3	1.8	1.9	NA	0	1.8
Neuronal and mixed neuronal glial	7.6	6.2	3.2	<2	3.1	0	1.3	0	2.4
MB and PNETs	13.9	19.8	25.7	15.4	14.6	17	28.9	10	22.4
Meningeal	3	2.6	1.2	<2	3.1	1.6	2.2	1.9	3.2
Nerve sheath	NA	0.4	NA	3.1	2.8	1.1	NA	0	3.6
Germ cell	3.6	8.1	NA	3.1	7.9	1.5	0.9	14.3	2
Craniopharyngioma	11	9.2	4.4	6.8	18.4	4.6	6.6	10.5	10.2
Pineal tumors	NA	NA	1.3	0.5	0.6	2.7	0.7	0	1.3

# Symptoms according to the tumor location

Anatomic Location	Common Signs and Symptoms
Frontal lobe	Personality changes, decreased motor speech (Broca's), seizures
Temporal lobe	Seizures, poor memory, language comprehension (Wernicke's)
Parietal lobe	Decreased sense of touch/pain, poor spatial and visual perception, poor interpretation of language
Occipital lobe	Poor/Loss of vision
Cerebellum	Ataxia, muscle movement/coordination, posture
Brainstem	Weakness, cranial neuropathies (III-XII), autonomic dysfunction
Thalamus	Weakness/motor control, consciousness, sleep/wake cycle
Hypothalamus	Autonomic dysfunction (temperature regulation, thirst, hunger, <i>etc.</i> ) endocrinopathies



	<b>Increased Intracranial</b>		
Generalized "Non-Localizing"	Pressure/Obstructive		
<u>Symptoms</u>	<u>Hydrocephalus</u>	"Localizing Symptoms"	Endocrine Symptoms
		Seizures (temporal or frontal	
Developmental Delay	Headache	lobe tumor)	Diabetes Insipidus
		Vision Changes (optic	
		pathway	
Behavioral Changes	Emesis	or occipital lobe tumor)	Hypothyroidism
		Motor Weakness (tumor in	
Decline in School Performance	Sleepiness/Lethargy	motor strip of cerebrum)	Weight Gain or Loss
	Papilledema (Vision	Cranial Neuropathies	
Tiredness/Sleepiness	Changes)	(brainstem tumor)	Panhypopituitarism
	Full/Bulging Fontanelle		Precocious Puberty

# Risk factors

- Radiation
- Genetics
- Head trauma
- Cell phones
- Diet, tobacco, Alcohol, Environment
- Infection/Immune system
- Allergies

# Radiation

- High dose radiation
- Treatment of benign conditions/ prophylactic cranial RT
- Meningiomas or GBM
- Radiation induced tumors mostly malignant

## Genetics

- Only 5% of brain tumors
- Familial aggregation non hereditary
- Increased incidence in first-degree relatives
- A small male gender predominance.
- "Glioma families" pattern of inheritance remains a mystery, revealing skipped generations

and inconstant times of onset.

(potential genetic etiologies or environmental risk factors - conflicting)

# NF 1 & 2

- 1 in 3000
- Chr 17
- Neurofibromin-restricts cell proliferation by activating (GTP) hydrolysis on ras proteins.
- Multiple Neurofibromas
- Fibrosarcomas, schwannomas, lowgrade astrocytomas - optic pathways, hypothalamus, cerebellum.

- NF2 gene mutation- TS gene AD
- Chr 22 membrane cytoskeletal protein
- Merlin/ Schwannomin.
- Bilateral vestibular schwannomas.
- Anaplastic or atypical meningiomas
- Other sporadic tumors

Von Hippel-Lindau syndrome.

### Li Fraumeni syndrome

- AD
- Chromosome 3p25 -tumor suppressor gene.
- Hemangioblastomas
- Pancreatic cysts
- Neuroendocrine tumors (including pheochromocytomas)
- Renal tumors.

- AD
- Chromosome TP53 –germline mutation
- Sarcomas
- Breast cancer
- Leukemia
- Adrenocortical cancer
- all occurring before the age of 45
- Brain tumors choroid plexus carcinomas.

### Tru cot syndrome.

Blue cell nevus syndrome

- An association between brain tumors and two forms of colonic polyposis – HNPCC and FAP
- HNPCC germline mutations MMR
- FAP -AD mutation APC gene Chr 5
- Majority Medulloblastomas,
   Gliomas

- Gorlin syndrome
- Increased risk of Medulloblastoma.
- Germline mutations
- Patched 1 (PTCH1) gene
- Tumor suppressorgene.

## TABLE 1-1 Hereditable Genetic Conditions

Condition	Gene Affected	CNS Lesion	Chromosome
Li-Fraumeni syndrome	TP53	Malignant glioma	17q
Neurofibromatosis type 1	NF1	Glioma of optic pathway/brainstem	17q11
Neurofibromatosis type 2	NF2	Acoustic neuroma, meningioma	22q12
von Hippel-Lindau syndrome	VHL	Hemangioblastoma of cerebellum/spinal cord	3p25
Turcot syndrome	APC	Glioblastoma, medulloblastoma	5q21
Gorlin syndrome	PTCH1	Medulloblastoma	9q22.3
<i>TP53</i> = tumor protein p53; <i>NF1</i> = neurofibromin 1; <i>NF2</i> = neurofibromin 2; <i>VHL</i> = von Hippel-Lindau			

tumor suppressor, E3 ubiquitin protein ligase; APC = adenomatous polyposis coli; PTCH1 = patched 1.

Original Article Published: 17 February 2017

Mobile phone use and risk of brain tumours: a systematic review of association between study quality, source of funding, and research outcomes

Manya Prasad <sup>[]</sup>, <u>Prachi Kathuria</u>, <u>Pallavi Nair</u>, <u>Amit Kumar</u> & <u>Kameshwar Prasad</u>

Neurological Sciences 38, 797–810 (2017) Cite this article

Letter to the Editor | Published: 08 July 2017

Use of cell phones and brain tumors: a true association?

<u>S. A. R. Mortazavi</u>, <u>Ghazal Mortazavi</u> & <u>S. M. J. Mortazavi</u>

Neurological Sciences 38, 2059–2060 (2017) Cite this article

Probabilistic Multiple-Bias Modeling Applied to the Canadian Data From the Interphone Study of Mobile Phone Use and Risk of Glioma, Meningioma, Acoustic Neuroma, and Parotid Gland Tumors @

F. Momoli ⊠, J. Siemiatycki, M. L. McBride, M.-É. Parent, L. Richardson, D. Bedard, R. Platt, M. Vrijheid, E. Cardis, D. Krewski

*American Journal of Epidemiology*, Volume 186, Issue 7, 1 October 2017, Pages 885–893, https://doi.org/10.1093/aje/kwx157

### Brain tumour risk in relation to mobile telephone use: results of the INTERPHONE international case-control study @

The INTERPHONE Study Group Author Notes

*International Journal of Epidemiology*, Volume 39, Issue 3, June 2010, Pages 675–694, https://doi.org/10.1093/ije/dyq079

#### Vol. 119, No. 11 | Commentary

#### Mobile Phones, Brain Tumors, and the Interphone Study: Where Are We Now?

Anthony J. Swerdlow , Maria Feychting, Adele C. Green, Leeka Kheifets, David A. Savitz, and International Commission for Non-Ionizing Radiation Protection Standing Committee on Epidemiology

Published: 1 November 2011 https://doi.org/10.1289/ehp.1103693 Cited by: 67

#### RESEARCH ARTICLE

Mobile phone use and glioma risk: A systematic review and meta-analysis

Ming Yang<sup>1</sup><sup>©</sup>, WenWen Guo<sup>2</sup><sup>©</sup>, ChunSheng Yang<sup>3</sup><sup>©</sup>, JianQin Tang<sup>4</sup>, Qian Huang<sup>2</sup>, ShouXin Feng<sup>1</sup>\*, AiJun Jiang<sup>1</sup>, XiFeng Xu<sup>1</sup>, Guan Jiang<sup>4</sup>\*

PLOS ONE | https://doi.org/10.1371/journal.pone.0175136 May 4, 2017

The World Health Organization (WHO) classified radiofrequency electromagnetic fields, such as those emitted by wireless phones, as "possibly carcinogenic to humans" (Group 2B) based on limited clinical evidence.



• Farmers and petrochemical workers

# Infections

- Viruses that induces genetic changes in cell
- Associations of infection with brain tumor inconsistent.
- polio vaccines contaminated with simian virus 40 (SV40) brain tumors
- Viral antigens from JC virus and human herpes virus 6 were detected in brain tumor subtypes but etiologic role is unclear
- Nucleic acids and proteins from CMV in GBM

## Allergy

#### Original paper | Published: 27 February 2013

Allergy and brain tumors in the INTERPHONE study: pooled results from Australia, Canada, France, Israel, and New Zealand

<u>Michelle C. Turner</u> <sup>⊡</sup>, <u>Daniel Krewski</u>, <u>Bruce K. Armstrong</u>, <u>Angela Chetrit</u>, <u>Graham G. Giles</u>, <u>Martine</u> <u>Hours</u>, <u>Mary L. McBride</u>, <u>Marie-Élise Parent</u>, <u>Siegal Sadetzki</u>, <u>Jack Siemiatycki</u>, <u>Alistair Woodward</u> & <u>Elisabeth Cardis</u>

Cancer Causes & Control 24, 949–960 (2013) Cite this article

- Interesting inverse relationship
- Prior varicella Zoster infection decrease risk of gliomas
- decreased incidence of glioma with a history of allergic sensitivity (asthma, eczema, or hayfever)
- ? Increased immune surveillance

#### Results

A significant inverse association was observed between a history of any allergy and glioma (OR = 0.73, 95 % CI 0.60–0.88), meningioma (OR = 0.77, 95 % CI 0.63–0.93), and acoustic neuroma (OR = 0.64, 95 % CI 0.49–0.83). Inverse associations were also observed with specific allergic conditions. However, inverse associations with asthma and hay fever strengthened with increasing age of allergy onset and weakened with longer time since onset. No overall association was observed for parotid gland tumors (OR = 1.21, 95 % CI 0.73–2.02).

# Interesting associations

- Physical activity (at least 51.6 MET-h of vigorous, moderate, and/or light intensity activity per week)
- Aged 15 and 18 had reduced glioma risk (RR=0.64; 95% CI=0.44-0.93; P trend =0.02)
- Aged 19-29 reduced riskf but not significant
- Aged 35-39 no association with the risk
- Height:  $\geq 1.9$  m have RR of 2.12
- Adolescent Obesity( at 18 years):  $BMI \ge 35$  (grade 3) has RR of 4.05
- Conflicting results...

Steven C. Moore *Cancer Res.* 2009 November 1; 69(21): 8349–8355. doi:10.1158/0008-5472.CAN-09-1669. Height, Body Mass Index, and Physical Activity in Relation to Glioma Risk.

# Contd...

- Head Truama: no associations; studies with positive results -? Recall bias
- Diet, vitamins, alcohol, tobacco, and environmental exposures Nitrate exposure from cured meats, tobacco, pesticides, synthetic rubber, vinyl chloride, or petrochemicals no significant associations
- Alcohol, antioxidants, fruits, vegetables reducing risk conflicting results

**Review Article** CONTINUUM

## Epidemiology and Diagnosis of Brain Tumors

Nicholas A. Butowski, MD

Address correspondence to Dr Nicholas A. Butowski, University of California, San Francisco, 400 Parnassus Avenue #0372, San Francisco, CA 94143, *butouski@neurosurg.ucsf.edu* **Relationship Disclosure:** Dr Butowski has served

Relationship Disclosure: Dr Butowski has served as a consultant for Prothena Corporation PLC and Proximagen and has received



#### Meta-Analysis

Birth Weight and Subsequent Risk of Childhood Primary Brain Tumors: A Meta-Analysis

- High birth weight associated with 2 MC brain tumors Astrocytoma and MB/PNET.
- Higher the weight higher the risk
- 1. High birth weight greater number of cells -in more cell divisions vulnerability to carcinogens.
- 2. Heuch et al. (for MB)- excess prenatal nutrition interfere with the migration of granular neuronal cells (30 weeks' gestation) Incompletely migrated cells immature increased neoplastic potential.
- 3. IGF-1 key role in brain ontogenesis, carcinogenesis;

- also in increased birthweight (maternal diabetes)

- Astrocytoma and MB

Rooth G. Increase in birthweight: a unique biological event and an obstetrical problem. Eur J Obstet Gynecol Reprod Biol

2003;106:86–7.

Bergmann RL, Richter R, Bergmann KE, et al. Secular trends in neonatal macrosomia in Berlin: influences of potential

# Spinal tumors

- 3% of CNS tumors and 4% in children 0.7 1.6 per 1,00,000
- Adult: >2/3 : nonmalignant.
- Childhood: Less frequent, more likely malignant; MC : Ependymoma
- Males MC: NST, ependymomas, astrocytomas; Females: Meningiomas
- Non Hispanic whites> Hispanics> non Hispanic blacks
- Mean age: 49 51 years

Descriptive epidemiology of primary spinal cord tumors. Kate A. Schellinger. J; Neurooncol (2008) 87:173–179 DOI 10.1007/s11060-007-9507-z

# Spinal tumors

WHO classification

- Extradural Secondaries (MC)
  - Primary benign: osteoma, osteoblastoma, ABC

Primary malignant: Myeloma/Plasmoacytoma, Chordoma, Chondrosarcoma,
 Osteosarcoma, Ewing's sarcoma

- Intradural Extramedullary(70 80%) NST, Meningioma, Ependymoma
  - Intramedullary(20-30%) Astrocytoma, Epedymoma,

Oligodendroglioma

- epidural hemangiomas, lipomas, extradural meningiomas, NST, lymphomas

- Indtradural: thoracic >cervical > lumbar



### TABLE 40.1 Primary Spinal Canal Tumors of Adults and Children: Types, Locations, and Frequencies

	Туре	Location	Frequency (%)
Adults	Meningioma	Intradural–extramedullary	37.6
	Nerve sheath tumor	Intradural–extramedullary	23.1
	Ependymoma	Intramedullary or intradural–extramedullary	20.5
	Astrocytoma	Intramedullary	4.2
	Hemangioma	Intramedullary	3.8
	Oligodendroglioma	Intramedullary	2.5
	Others	—	8.4
Children	Astrocytoma	Intramedullary	31.4
	Ependymoma	Intramedullary or intradural–extramedullary	21.6
	Oligodendroglioma	Intramedullary	16.8
	Nerve sheath tumor	Intradural–extramedullary	13.3

## Intramedullary Spinal Cord Tumors: Part I— Epidemiology, Pathophysiology, and Diagnosis

Dino Samartzis<sup>1,2</sup> Christopher C. Gillis<sup>3</sup> Patrick Shih<sup>4</sup> John E. O'Toole<sup>3</sup> Richard G. Fessler<sup>3</sup>

Tumor class	Associated tumors
Neuroepithelial tissue	Astrocytic • Astrocytoma • Glioblastoma Embryonal • Primitive neuroectodermal Ependymal • Ependymoma • Subependymoma Mixed glioma Neuronal and mixed neuronal-glial • Gangliocytoma • Ganglioglioma • Ganglioneuroblastoma Oligodendroglial • Oligodendroglioma Uncertain origin • Polar spongioblastoma
Spinal nerves	Neurofibroma Schwannoma
Nonmeningothelial, mesenchymal	Hemangioblastoma Lipoma Melanoma Sarcoma
Germ cell tumors	Germinoma Teratoma
Cysts and tumorlike lesions	Dermoid Epidermoid
Hematopoietic neoplasms	Primary central nervous system lymphoma (microglial)
Metastatic tumors and other rare neoplasms	

# Symptoms and signs

- Pain localized or radiating 72%
- Weakness 55%
- Sensory deficits 39%
- Sphincter dysfunction 15%
- Bladder and bowel dysfunction: conus medullaris and filum terminale

Cauda equina nerve root compression syndrome:

- Radicular pain anterior (L4), lateral (L5), or posterior (S1) thigh
- Muscle wasting glutei, hamstrings, or tibialis anterior
- Saddle anaesthesia
- Absent ankle reflexes (S1), or plantar (S2) responses
- Impotence
- Loss of anal or bulbar cavernous reflexes

# Risk factors

### NF1, NF2, VHL

Genetic disorder	Associated tumor
Neurofibromatosis-1	Acute nonlymphocytic leukemiasAstrocytomasCarcinoid tumorsHematomasHypothalamic gliomasMalignant nerve sheath tumorsMeningiomasNeurofibromasOptic nerve gliomasPheochromocytomasPrimitive neuroectodermal tumorsRhabdomyosarcomasWilms tumor
Neurofibromatosis-2	Bilateral acoustic schwannomas Ependymomas Gliomas Meningiomas Schwannomas
Von Hippel-Landau	Adrenal pheochromocytomas Central nervous system hemangioblastoma (medulla oblongata or spinal cord) Epididymal cystadenomas Lindau tumor (cerebellum hemangioblastoma) Nephritic cysts Pancreatic cysts Renal cell carcinomas Renal cysts Renal cysts Retinal hemangioblastomas



# Syringomyelia

- Fluid-filled cavities in the spinal cord
- 25 to 58% of patients with IMSCT
- MC Lower Cervical and Upper Thoracic region
- MC Ependymomas, Hemangioblastomas, Cavernomas
- Degree of cephalad extension for a tumor and the presence of a syrinx correlated
- 49% above, 11% below tumor level, and 40% are bipolar
- Favorable prognostic sign Noninfiltrative tumors with distinct cleavage planes than more diffuse, infiltrative tumors
- Typically resolves with tumor removal

Intramedullary Spinal Cord Tumors: Part I— Epidemiology, Pathophysiology, and Diagnosis. Dino Samartzis. Global Spine J 2015;5:425–435

# Syringomyelia – classification and etiology

(1) Alteration of cerebrospinal fluid (CSF) flow dynamics related to hindbrain disorders

(2) Intramedullary tissue damage secondary to hemorrhage or infarction – MC with Ependymomas, Hemangioblastomas, Cavernomas

(3) resulting from direct secretory ability of intramedullary tumor – increased protein levels vs CSF and non IMSCT syrinx

- DD: CSF obstruction and hindbrain disorders
  - Gardner's "water hammer" theory
  - Williams' "suck and slosh" theory
  - Ball and Dayan's theory of infiltration through perivascular (Virchow-Robbin) spaces
  - Oldfield's theory, which combines features of all of the above

Intramedullary Spinal Cord Tumors: Part I— Epidemiology, Pathophysiology, and Diagnosis. Dino Samartzis. Global Spine J 2015;5:425–435

# Summary

- Recognizing patterns of health and diseases
- <u>Understanding the variables that influence these patterns</u>
- Risk factors for their occurences
- Reasons for the outcomes



